

**AN UPDATE ON AETIOLOGY OF SUDDEN SENSORINEURAL  
HEARING LOSS IN ADULTS: A SYSTEMATIC REVIEW**

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**This Dissertation is submitted as part of  
fulfilment for the Degree of Master of Science in Audiology  
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**September 2021**

## **CERTIFICATE**

This is to certify that this dissertation entitled '**An Update on Aetiology of Sudden Sensorineural Hearing Loss in Adults: A Systematic Review**' is a bonafide work submitted as a part for the fulfilment for the degree of Master of Science (Audiology) of the student Registration Number: 19AUD012. This has been carried out under the guidance of the faculty of this institute and has not been submitted earlier to any other University for the award of any other Diploma or Degree.

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## **DECLARATION**

This is to certify that this dissertation entitled '**An Update on Aetiology of Sudden Sensorineural Hearing Loss in Adults: A Systematic Review**' is the result of my own study under the guidance of Dr K. Rajalakshmi, Department of Audiology, All India Institute of Speech and Hearing, Mysore and has not been submitted earlier to any other University for the award of any other Diploma or Degree.

Mysuru  
September 2021

**Registration Number: 19AUD012**

*Dedicated to Achan, Amma,  
Chikku & Lachu..*

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*I would like to thank all my friends, especially **Praveen**, who helped me to complete my dissertation I also thank all lecture and clinical staff who had improved my clinical as well as academic knowledge*

## **ABSTRACT**

The present review aimed to document the evidence for aetiology of sudden SNHL from literature published in the last ten years. A totally internet-based search was carried out for the literature in the database of Google Scholar, Pubmed, and J-GATE from December 2020 to July 2021. Articles regarding the topic of aetiology of sudden SNHL were searched in the mentioned database. The articles published from 2011 to 2021 July were considered to select for the review. The data extraction were performed by Rayyan intelligent, systematic review software developed by Qatar Computing Research Institute (QCRI). Based on the inclusion and exclusion criteria, 7 articles were selected for systematic review from 1,943 articles obtained after the initial search. The NIH study quality assessment tool was used for the quality analysis of finalized articles.

The summary of all 7 selected articles identified idiopathic as a major cause for sudden SNHL and other causes reported in the reviewed articles are as follows, inner ear or intra labyrinthine haemorrhage, acoustic trauma, head injury, vestibular schwannoma or acoustic neuroma, meningioma, labyrinthitis, structural deformity, perilymphatic fistula and systemic autoimmune diseases such as Lupus erythematosus, rheumatoid arthritis, scleroderma, vitiligo, Cogan's syndrome, psoriasis, Susac syndrome, Sjogren syndrome and ankylosing spondylitis. The updated articles in the current study may help the audiologist perform appropriate diagnostic test batteries to find the causal link between the various aetiological factors and sudden SNHL.

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## Chapter 1

### INTRODUCTION

Hearing loss is the partial or total inability to hear sounds in one or both ears, which is considered to be one of the most common sensory deficits in human beings. It can occur due to congenital (present since birth) or acquired reasons. Hearing deficit can be classified based on its sites of lesions, such as conductive (external or middle ear dysfunction, or both), sensorineural (inner ear dysfunction) and mixed type (dysfunction or external ear and or middle ear and inner ear). Conductive hearing loss can be due to congenital malformation of the external or middle ear such as atresia, anotia, microtia (external ear), fused middle ear ossicles, absence of stapedius tendon (middle ear) or acquired causes such as infections of the external ear or middle ear, foreign body obstruction of the ear canal, etc. The aetiology of sensorineural hearing loss can be congenital, such as genetics, lack of oxygen during birth, infectious diseases passed from mother during pregnancy period (eg. Rubella), or acquired such as presbycusis, induced by noise, induced by drugs, viral infections etc. The common causes of mixed hearing loss are the combination of congenital malformation in the external, middle and inner ear or acquired infections such as chronic otitis media, cholesteotoma, etc.

Onset of Hearing loss can be sudden or progressive; hence, whenever the onset of the hearing loss is sudden and the site of lesion is in the inner ear, it is termed as sudden sensorineural hearing loss (SSNHL). There is no universally accepted definition for sudden sensorineural hearing loss. However, it is most often defined as a sensorineural hearing loss of 30 dB or greater in at least three continuous audiometric frequencies occurring over 72 hours (Wilson, Byl, & Laird, 1980).

Despite comprehensive research, uncertainty exists in the aetiology and proper treatment of patients with idiopathic sudden SNHL. However, the causes of sudden SNHL are broadly classified into two, idiopathic (unknown cause) and identifiable cause. The majority of patients with sudden SNHL are listed as 'idiopathic' and have no known cause of hearing loss, although identifiable causes are found for 7% to 45% of patients with sudden SNHL (Shaia & Sheehy, 1976; Byl, 1984; Fetterman et al., 1996; Chau et al., 2010). Due to the enormous range of causes, only ten percent of people diagnosed with SSNHL appear with identifiable aetiologies. Some of them are infections, head trauma, autoimmune disorders, drug consumption for severe infections, tumors and cancer, vascular pathologies, neurological disorders and other inner ear pathologies. Hence, most of the cases are accompanied by any other medical conditions (NIDCD, 2018). Hence an appropriate assessment and treatment of sudden SNHL remain controversial, with over 100 aetiologies for this syndrome currently being suggested (Jaffe, 1973). Stokroos & Albers (1996) conducted a literature review to assess the supporting evidence for the different theories of aetiology of idiopathic sudden SNHL and found the most needed evidence of the four common theories such as impairment of labyrinthine blood supply, viral infection of the labyrinth, inner ear membrane ruptures, and autoimmune mediated cochlear malfunction. Chau et al (2010) reviewed 23 studies of sudden SNHL. They found that the suspected aetiology of sudden SNHL were idiopathic or identifiable causes such as infectious disease followed by otologic disease, trauma, vascular, neoplastic, and other causes.

Epidemiological data reveals the incidence of sudden SNHL range from 5 to 20 per 100,000 persons per year and any age group may be affected. However, incidence found to be in 5<sup>th</sup> Or 6<sup>th</sup> decade of life (Byl, 1984). In United States the incidence of sudden SNHL has been found to be 27 per 100,000 during 2006 and 2007

with a male-to-female ratio of 1.07:1; there was a little male preponderance. The incidence was more noticed in people aged 65 and older (Alexander, Thomas, Harris and Jeffrey, 2013)

Regardless of aetiology, there may be no, partial or full recovery of hearing thresholds following sudden SNHL. The recovery rate depends on many factors such as age at the onset of hearing loss, degree of hearing loss and impaired frequencies, the occurrence of vertigo, and time between the onset of hearing loss and the visit to the treating doctor (Byl, 1984). A sudden, stable hearing loss is noted by some patients, while others experience increasingly progressive loss. Any audiometric frequency range can be involved in hearing loss. Associated tinnitus, vertigo, and aural fullness have been described (Byl, 1994; Einer et al.,1994; Nosrati-Zarenou et al., 2007).

The management of sudden SNHL is with the help of corticosteroids, especially when the hearing loss is due to idiopathic causes. In earlier times, the medication was given in the pill form, whereas currently, it can be given as intratympanic (through the eardrum) injections. Literature also revealed that intratympanic injections are efficient than oral consumption. Medication has to be given as early as possible otherwise, the rate of recovery can be affected. Additional medications can be given for the accompanying medical conditions. If hearing loss is not reversible in nature, other amplification devices such as hearing aids or cochlear implants can be recommended.

## **1.1 Need for the study**

From the above literature, it is clear that a broad range of aetiological conditions can cause sudden SNHL. Hence, it is very difficult to directly link sudden SNHL and these various aetiology (Chau et al., 2010). Hellmann et al. (2011) found sudden SNHL as an early sign of multiple sclerosis. Daniel et al. (1989) also found sudden SNHL as a presentation of multiple sclerosis.

Jeong et al. (2019) carried out a study to see the association of sudden SNHL and several autoimmune diseases and found the risk of sudden SNHL significantly higher in patients with autoimmune diseases such as rheumatoid arthritis, which may damage other parts of a patient's body. Charlene et al. (2013) done a population-based cohort study to find the correlation between sudden SNHL and acute myocardial infarction and found that sudden SNHL may advice an independent risk of myocardial infarction so this observation may help in early detection and proper treatment of patients with a high risk of myocardial infarction.

Knowledge of various aetiology of sudden SNHL may help the audiologist guide the patients for proper treatment and further risk factors associated with sudden SNHL. Hence, this review aims to document the evidence for aetiology of sudden SNHL from literature published in the last ten years.

## Chapter 2

### METHOD

A totally internet based search was carried out for the literature in the data base of Google Scholar, Pubmed and J-GATE from December 2020 to July 2021. Articles regarding the topic of aetiology of sudden SNHL were searched in the mentioned database. The articles published since 2011 to 2021 July in were considered to select for the review. To find more article an advanced search was done using BOOLEAN operations such as AND, OR and NOT with different key words relating to the study topic in all the data base. The combination of different key words used were as follows: (((("aetiologie"[All Fields] OR "aetiologies"[All Fields] OR "aetiology"[All Fields] OR "etiologies"[All Fields] OR "etiology"[MeSH Subheading] OR "etiology"[All Fields] OR "causality"[MeSH Terms] OR "causality"[All Fields]) AND ("sudden"[All Fields] OR "suddenness"[All Fields]) AND ("hearing loss, sensorineural"[MeSH Terms] OR ("hearing"[All Fields] AND "loss"[All Fields] AND "sensorineural"[All Fields]) OR "sensorineural hearing loss"[All Fields] OR ("sensorineural"[All Fields] AND "hearing"[All Fields] AND "loss"[All Fields]))) OR (("causative"[All Fields] OR "causatively"[All Fields] OR "causatives"[All Fields] OR "cause"[All Fields] OR "caused"[All Fields] OR "causing"[All Fields] OR "etiology"[MeSH Subheading] OR "etiology"[All Fields] OR "causes"[All Fields] OR "causality"[MeSH Terms] OR "causality"[All Fields]) AND ("hearing loss, sudden"[MeSH Terms] OR ("hearing"[All Fields] AND "loss"[All Fields] AND "sudden"[All Fields]) OR "sudden hearing loss"[All Fields] OR ("sudden"[All Fields] AND "hearing"[All Fields] AND "loss"[All Fields]))) OR (("aetiologie"[All Fields] OR "aetiologies"[All Fields] OR "aetiology"[All Fields] OR "etiologies"[All Fields] OR "etiology"[MeSH Subheading] OR "etiology"[All Fields] OR "causality"[MeSH

Terms] OR "causality"[All Fields]) AND ("hearing loss, sudden"[MeSH Terms] OR ("hearing"[All Fields] AND "loss"[All Fields] AND "sudden"[All Fields]) OR "sudden hearing loss"[All Fields] OR ("sudden"[All Fields] AND "deafness"[All Fields]) OR "sudden deafness"[All Fields])) AND (y\_10[Filter]). Once all the advanced search were completed the selection of articles for the study was done with a predetermined inclusion and exclusion criteria which are as follows.

### **Inclusion criteria**

- Studies published since 2011
- Retrospective reviews of consecutive patients with sudden SNHL
- Prospective cohort studies in sudden SNHL
- Studies with randomized controlled trials of patents with sudden SNHL
- Studies published in English language
- Studies on human subjects

### **Exclusion criteria**

- Non-consecutive prospective studies
- Non-consecutive retrospective studies
- Practice guidelines
- Studies include children as population
- Case studies in sudden SNHL
- Review articles

### **2.1 Data extraction**

The data extraction was performed by Rayyan intelligent, systematic review software developed by Qatar Computing Research Institute (QCRI). The review was



completed independently by two reviewers (Athul P R and Praveen Prakash). All the duplicates were removed by using Rayyan software, and after removing the duplicates, title screening was done by the reviewers independently. The titles which include keywords relevant to aetiology or various causes of sudden SNHL were considered for abstract screening. All the abstracts were then reviewed with predetermined inclusion and exclusion criteria. The abstracts of studies that fulfilled the inclusion criteria were retrieved for full-text screening. The full-text screening and quality analysis were carried out independently by each reviewer and finalized the articles to summarize for the review. PRISMA guidelines were followed for the screening and finalizing the articles for review.

## **2.2 Quality analysis (Risk of bias)**

The quality analysis was performed for the finalized articles by two reviewers to avoid bias. The NIH study quality assessment tool was used for the quality analysis. The NIH study quality assessment tool had a different questionnaire for Observational cohort and cross-sectional studies, Before and after (pre-post) studies with no control group and case series studies. Each of these mentioned tools had a different number of questions fourteen, twelve and nine, respectively, regarding the study to rate the quality of articles. The questions were provided with yes/no/other options to be answered reading each article. Based on the study type mentioned in the article, the assessment tool was selected for the quality analysis. The overall quality of the study was rated as Good, Fair or Poor based on the yes/no response of the questions regarding the study. Two reviewers had performed the quality analysis independently and rated the articles as Good, Fair or Poor. It was pre-planned to approach a third reviewer if the quality analysis had any discrepancy; since there was not much significant difference between the initial two reviewers' rating, the third review for

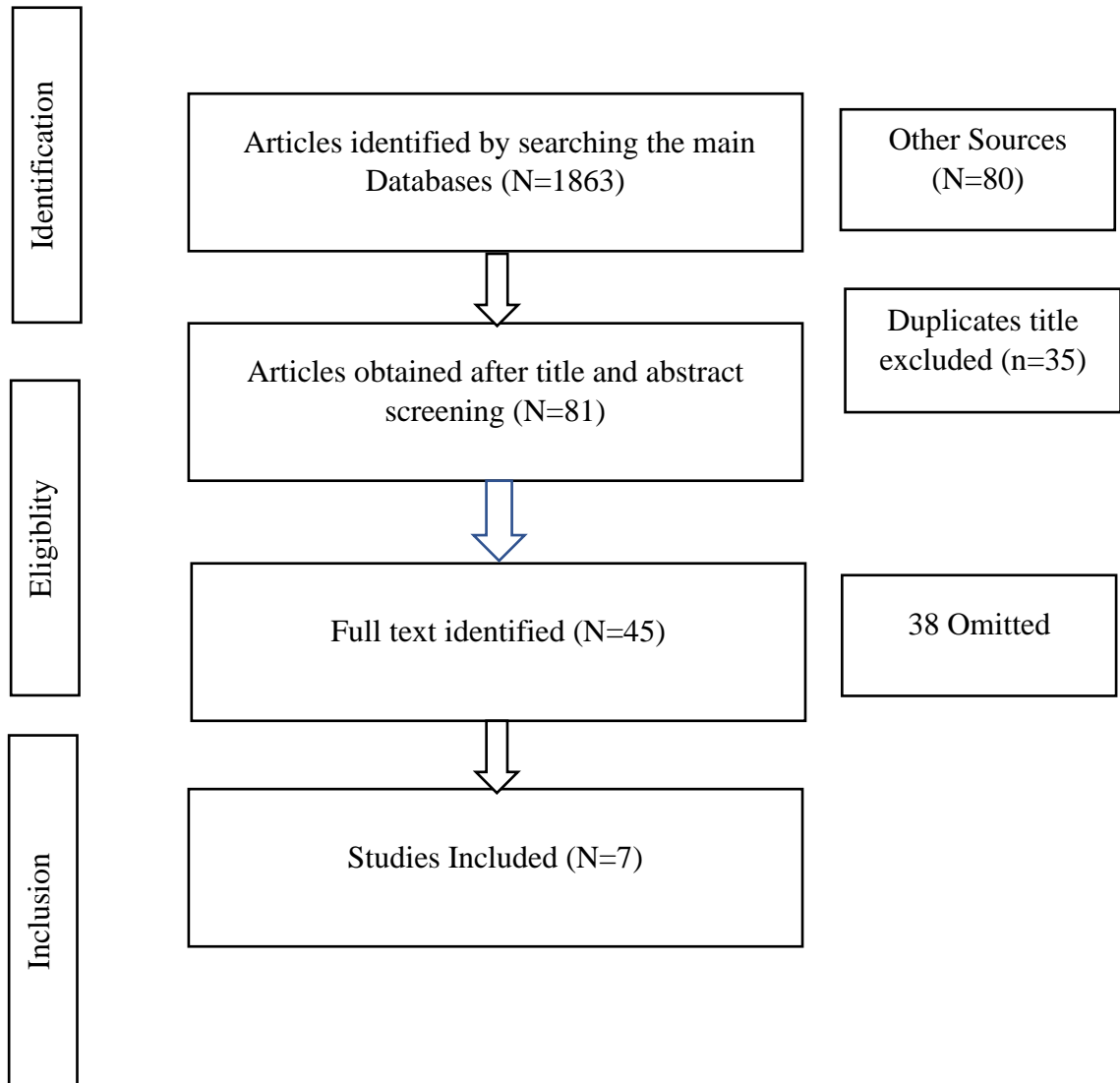
quality analysis was not carried out. Examples for some of the questions in the quality assessment tool are as follows: “*Were eligibility/selection criteria for the study population prespecified and clearly described?*”, “*Were all eligible participants that met the prespecified entry criteria enrolled?*”, “*Was the sample size sufficiently large to provide confidence in the findings?*”, “*Were all the subjects selected or recruited from the same or similar populations (including the same time-period)? Were inclusion and exclusion criteria for being in the study prespecified and applied uniformly to all participants?*”, “*Were the exposure measures (independent variables) clearly defined, valid, reliable, and implemented consistently across all study participants?*”, “*Were the results well-described?*” etc.

## **Chapter 3**

### **RESULTS**

A total of 1,943 studies were obtained after conducting all the searches from the three different databases which is mentioned in the method part. After removing 35 duplicates, 1,908 articles underwent title and abstract screening, and 45 articles were finally selected for full-text screening. Based on the inclusion and exclusion criteria, 7 articles were finalized for performing the review. The quality analysis was done by two reviewers independently and three articles were rated as ‘Good’ and the remaining four articles were rated as fair. No article was rejected by conducting quality analysis. Figure 3.1 illustrates the summary of the article selection procedure, starting from literature search results from a different database to finalizing the article for systematic review. Table 3.1 represents the demographics of the results of quality analysis done by using the NIH study quality assessment tool for the articles for review. Table 3.2 depicts the summary of selected studies for the review, which listed the various aetiology of sudden SNHL.

All seven finalized articles listed one or more causes of sudden SNHL. Various diagnostic tests were performed to conclude the precise cause of sudden SNHL for the study population. Out of 7 studies, only one had included the study population with single aetiology of sudden SNHL (Chen et al., 2018). All other 6 studies had a minimum of two aetiologies reported to cause sudden SNHL.



**Figure 3.1:** PRISMA chart showing the summary of the article selection procedure.

SL No	Author	Study Type	Year	Country	Number Of Total Participants	Quality Analysis
1.	Chen et al	Retrospective	2018	China	42	Good
2.	Gupta et al	Prospective	2016	India	37	Fair
3.	Cho et al	Retrospective	2017	Korea	200	Good
4.	Rossini et al	Retrospective cohort	2016	Brazil	13	Fair
5.	Lee et al	Retrospective	2016	Korea	35	Fair
6.	Haubner et al	Retrospective	2012	Germany	69	Fair
7.	Hosokwa et al	Retrospective	2018	Japan	104	Good

**Table 3.1:** Demographics of articles and results of quality analysis done by NIH study quality assessment tool

Author	Age Range	Number of total participants	Tests Performed	Etiology	Results	Implication
Chen et al	39.3 ± 14.8 Years	42	Pure tone audiometry (PTA), Word Recognition Score (WRS), and Magnetic Resonance Imaging (MRI)	Inner Ear Hemorrhage (IEH)	All SSNHL patients were positive for IEH in MRI. Intravenous (IV) Methylprednisolone alone was used for 24 patients and both Intravenous Methylprednisolone and intratympanic (IT) corticosteroid were used for 18 patients in the treatment. The mean PTA and WRS at 6 months were 90.8 ± 16.2 dB and 38.7 ± 35.5 percent,	Results implied that immediate and effective intratympanic corticosteroid might be a potential remedy for IEH.

					<p>respectively, which were statistically better than at baseline. (<math>109.2 \pm 9.6</math> dB, <math>p = 0.000</math>; and <math>1.3 \pm 4.0\%</math>, <math>p = 0.000</math>). The patients who were treated with both Intravenous Methylprednisolone and intratympanic (IT) corticosteroid showed more improvement in WRS after 6 months.</p>	
Gupta et al	>18 Years	37	ENT examination, Pure tone audiometry,	Idiopathic (24), Acoustic trauma	CT scan of head in patients with head injury indicated extradural hemorrhage.	MRI should be performed in all patients with

			Impedance audiometry, MRI, and Computed tomography (CT) scan	(9) and Head injury (4)	Intravenous methylprednisolone, tab. prednisolone, tab. pentoxifylline and injection of methylcobalamin intramuscularly were used for treatment. Out of 24 patients with idiopathic sudden SNHL 11, 7 and 6 achieved complete, partial, and no recovery. 6 and 3 patients achieved partial and no recovery among 9 patients with acoustic trauma associated with	sudden SNHL, irrespective of recovery in hearing loss. Associated symptoms like vertigo and comorbidities such as hypertension and diabetes mellitus along with the flat configuration of audiogram, are poor prognostic
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					sudden SNHL. In patients with head injury associated with sudden SNHL, two patients each achieved partial and complete recovery.	factors in the recovery of hearing loss.
Cho et al	18 to 84 Years	200	Pure tone audiometry, Spontaneous nystagmus (SN), Head-shaking nystagmus(HSN), Positional nystagmus (PN), Bithermal caloric	Idiopathic(175), Vestibular schwannoma(9), Meningioma(1), Intra-labyrinthine haemorrhage (ILH 6), Labyrinthitis(7)	Among 200 patients, 25 had abnormal MRI findings indicating inner ear lesions hence they were considered as secondary group and the rest 175 patients were considered as idiopathic group. Vertigo was present in six of seven patients with	The secondary SSNHL group was associated with a poor prognosis, in which ILH and labyrinthitis were the most common findings (6.5%), and their

			<p>test (neuro-otological tests).</p> <p>Lipd battery,</p> <p>Thyroid function and serological tests (HBsAg, Anti HBs, Anti HCV, HIV).</p> <p>MRI</p>	<p>and Structural deformity(2)</p>	<p>ILH, and in 3 of 6 patients with labyrinthitis, but none among the IAC tumor group had no complaint of vertigo. Patients with IAC tumors did not correspond to those with BPPV, whereas three patients in ILH group and two in the labyrinthitis group were diagnosed as BPPV. Hence the presence of vertigo and BPPV in the tumorous and non-tumorous groups was significantly different. A number of</p>	<p>clinical characteristics also differed from those of the VS group as well as those from the idiopathic SSNHL group. MRI evaluation of patients with SSNHL for IAC tumors cannot be omitted, even in patients with complete hearing recovery.</p>
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					patients with complete recovery was more in the idiopathic group compared to the second group in propensity score-matched groups. One patient with vestibular schwannoma had complete recovery after treatment	
Rossini et al	21 to 68 Years	13	Pure tone audiometry, Recognition threshold for speech and Speech recognition index.	Systemic autoimmune diseases- Systemic Lupus Erythematosus	All 13 patients had normal external auditory canal (EAC) and tympanic membrane (TM) examined by otoscope. MRI was done for 10 patients (76.92%)	The patients with sudden SNHL and SAD have more severe impairment initially, more bilateral

			<p>Blood tests which include complete blood count, lipid dosage, serum fasting glucose, renal function and thyroid function erythrocyte sedimentation rate and serology for syphilis, AIDS, borreliosis (when necessary).</p> <p>MRI. Patients suspected systemic</p>	<p>(SLD-4), Rheumatoid Arthritis (RA-2) Scleroderma + RA (1), RA + Vitiligo (1), Cogan's syndrome (1), Psoriasis(1), Susac syndrome(1), Sjogren syndrome +Sclroderma (1)</p>	<p>where none of them showed cochlear enhancement on T1- weighted contrast injection. Corticosteroid prednisone and immunosuppressants were used for treatment in all patients. Out of 13 patients, 3 had sudden bilateral SNHL. Among them, one had simultaneous involvement with SLE, two had contralateral posterior involvement 15 days after the first attack, and the other</p>	<p>involvement, poor response to treatment, and worse prognosis compared to patients with sudden SNHL of unknown etiology.</p>
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			<p>autoimmune diseases (SAD) underwent some specific serological tests.</p>	<p>and Ankylosing spondylitis</p>	<p>had underlying disease as Susac syndrome The other had Cogan's syndrome.</p>	
Lee et al	19 to 71 Years	35	<p>Pure tone audiometry, MRI, lab tests for hemoglobin, WBC, platelet, PT, PTT</p>	<p>Idiopathic(23) and Intralabyrinthine hemorrhage(ILH-12)</p>	<p>The majority of patients in the hemorrhage group (92%) complained of dizziness even though the frequency of lesions on the vestibular structure was not correlated with this symptom. The initial degree of hearing impairment in the hemorrhage group was</p>	<p>ILH has an association with poor hearing prognosis and the occurrence of vertigo in sudden SNHL. T1-weighted MRI can help to understand the etiology of</p>

					higher than the idiopathic group (94±35.9 vs 66±30.1). The final hearing threshold in the hemorrhage group was worse (78.19±46.26 vs 37±31.96). The hearing recovery was more at low frequency than high frequency in the hemorrhage group	sudden SNHL in patients with intralabyrinthine hemorrhage.
Haubner et al	18 to 92 Years	69	Pure tone audiometry and Exploratory tympanotomy	Idiopathic (41) Perilymphatic fistula (definite-13, doubtful-15)	The exploratory tympanotomy of 41 patients suggested no fistula and considered them as an idiopathic group. A definite	A safe exploratory tympanotomy procedure along with sealing of round window

					<p>sign of round membrane rupture which indicates a clear perilymphatic fistula was found in 13 (18.8%) patients and a doubtful perilymphatic fistula in 15 (21.7%) patients. All the patients underwent an exploratory tympanotomy and received a sealing of the round window niche. In the majority of the patients, the postoperative PTA was significantly improved compared to preoperative</p>	<p>niche may result in hearing improvement in patients with sudden SNHL due to perilymphatic fistula along with established treatment regimen such as high dose steroid</p>
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					data. In the group of definite perilymphatic fistula, 7 patients (53%) had an improvement in hearing more than 20 dB after the tympanotomy.	
Hosokwa et al	43.1±11.8	104	Pure tone audiometry and MRI	Idiopathic (90) and Acoustic neuroma (14)	The mean PTA average for sudden SNHL idiopathic group and sudden SNHL acoustic neuroma group was 45.1±10.9 and 58.8±15.9. The severity of hearing loss was more in idiopathic group compared to acoustic neuroma group. All the 104	The mean hearing loss at 125 Hz, 250 Hz and 500 Hz was significantly milder in patients with sudden SNHL and acoustic neuroma who had a trough-shaped



					<p>participants with sudden SNHL had a trough shaped audiogram. 90 (10.9%) patients with trough shaped audiograms were selected from 828 patients with sudden SNHL and 14 (70.0%) were selected from 20 patients with sudden SNHL with acoustic neuroma. All the patients were treated with hydrocortisone 400 mg/kg, which was tapered over 10 days. The recovery rate</p>	<p>audiogram than in those with idiopathic sudden SNHL. The incidence of a trough-shaped audiogram is higher in patients with sudden SNHL and acoustic neuroma than in those with idiopathic sudden SNHL. A trough-shaped audiogram</p>
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					percentage was high in the idiopathic group compared to acoustic neuroma with sudden SNHL group. In patients with sudden SNHL and acoustic neuroma, the mean age was 44.7, less than patients with acoustic neuroma only. The mean tumor size was 7.25 mm in patients with sudden SNHL with acoustic neuroma and 18.7 mm in patients with only acoustic neuroma.	in patients with sudden SNHL, especially with minimal to mild low-frequency hearing loss, could indicate acoustic neuroma.
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**Table 3.2:** Summary of all final 7 studies listed with various aetiology of sudden SNHL. (**Abbreviations:** tab; tablet, WBC; white blood cells PT; prothrombin time, PTT; partial thromboplastin time)

## **Aetiology of sudden SNHL**

The majority of the study population in five of seven articles had idiopathic causes as aetiology for sudden SNHL (Gupta et al., 2016; Cho et al., 2017; Lee et al. et al., 2016; Haubner et al., 2012; Hosokwa et al., 2018). In three studies, inner ear or intra-labyrinthine hemorrhage has been confirmed as an aetiology of sudden SNHL for 42, 6 and 12 patients done by Chen et al. (2018), Cho et al. (2017) and Lee et al. (2016) respectively. Cho et al. (2017) and Hosokwa et al. (2018) could add 9 and 14 patients respectively with vestibular schwannoma as a leading cause for sudden SNHL. Other tumours such as meningioma can also cause sudden SNHL (Cho et al., 2017).

Rossini et al. (2016) described 13 patients with systemic autoimmune diseases (SAD) who had sudden SNHL due to the same such as Lupus Erythematosus (SLD-4), Rheumatoid Arthritis (RA-2) Scleroderma + RA (1), RA + Vitiligo (1), Cogan's syndrome (1), Psoriasis(1), Susac syndrome(1), Sjogren syndrome + Scleroderma (1) and Ankylosing spondylitis and recorded that the patients with sudden SNHL and SAD have more severe impairment initially, more bilateral involvement, poor response to treatment and worse prognosis compared to patients with sudden SNHL of unknown aetiology. Haubner et al (2012) done exploratory tympanotomy in 69 patients with sudden SNHL and found 41 patients had idiopathic aetiology for sudden SNHL and a definite sign of round membrane rupture, which indicate a clear perilymphatic fistula was found in 13 patients and a doubtful perilymphatic fistula in 15 patients. And he reported that a safe exploratory tympanotomy procedure along with sealing of round window niche might result in hearing improvement in patients with sudden SNHL due to perilymphatic fistula along with established treatment regimen such as high dose steroid.

Hosokwa et al. (2018) studied 104 sudden SNHL patients with trough-shaped audiograms where 90 patients had idiopathic cause and 14 patients had acoustic neuroma as an aetiology of sudden SNHL. They recorded that the mean hearing loss at 125 Hz, 250 Hz and 500 Hz was significantly milder in patients with sudden SNHL and acoustic neuroma. They had a trough-shaped audiogram than in those with idiopathic sudden SNHL. The incidence of a trough-shaped audiogram is higher in patients with sudden SNHL and acoustic neuroma than in those with idiopathic sudden SNHL. A trough-shaped audiogram in patients with sudden SNHL, especially with minimal to mild low-frequency hearing loss, could indicate acoustic neuroma. Table 3.3 listed the number of cases with specific aetiology in each study and the total number of patients per aetiology

<b>Aetiology</b>	<b>Author</b>	<b>Number of cases</b>	<b>Total</b>
Idiopathic	Guptha et al	24	353
	Cho et al	175	
	Lee et al	23	
	Haubner et al	41	
	Hosokwa et al	90	
Inner ear / intra-labyrinthine haemorrhage	Chen et al	42	60
	Cho et al	6	
	Lee et al	12	
Vestibular schwannoma/acoustic neuroma	Cho et al	9	23
	Hosokwa et al	14	
Systemic autoimmune disease	Rossini et al	13	13
Perilymphatic fistula	Haubner et al	28	28
Other aetiology	Gupta et al	13	23
	Cho et al	10	
<b>Total</b>			<b>500</b>

**Table 3.3:** List of number of cases with specific aetiology in each study and a total number of patients per aetiology.

## Chapter 4

### DISCUSSION

Sudden sensorineural hearing loss can be caused by a variety of factors, the most common of which is idiopathic. Apart from the idiopathic cause, a wide range of disorders of the auditory system can contribute to the development of sudden SNHL and these can be listed under major causes such as infectious causes, traumatic causes, neoplastic causes, immunological causes, circulatory causes, toxic causes, neurologic causes, metabolic causes and other cause like Meniere's disease, genetic predisposition, stress, neurosarcoidosis and pseudohypocacus (Hughes et al., 1996). A review done by Chau et al. (2011), it recorded as idiopathic was the primary cause and other factors listed under various diseases such as infectious diseases (Eg: adenovirus, hepatitis C, herpes simplex virus, measles, mumps, rubella, varicella-zoster, meningitis, syphilis etc.), otologic disease (Eg: Meniere's disease, hydrops, otosclerosis, autoimmune inner ear disease, skull base or otologic surgery and aminoglycoside toxicity), trauma (Eg: head injury or skull fracture, acoustic trauma, barotrauma and trauma after water irrigation), vascular/ hematologic (Eg: neurovascular conflict, cardiovascular disease, subdural hematoma, pontine hemorrhage, transient ischemic attack, sickle cell anemia, and hemodialysis coagulopathy), neoplastic (Eg: vestibular schwannoma, meningioma, cerebellar angioma and multiple myeloma) and other causes like other CNS disease, nonotologic surgery postoperative, pregnancy-related, post rabies vaccination and carbon monoxide poisoning. A recent review study showed a direct association between COVID-19 and sudden SNHL and the virus can affect the auditory system either directly or indirectly (Umashankar et al., 2021).

The present study has listed idiopathic as a major cause for sudden SNHL as previous literature and other causes reported in the reviewed articles are as follows, inner ear or intra-labyrinthine hemorrhage, acoustic trauma, head injury, vestibular schwannoma or acoustic neuroma, meningioma, labyrinthitis, structural deformity, perilymphatic fistula and systemic autoimmune diseases such as Lupus erythematosus, rheumatoid arthritis, scleroderma, vitiligo, Cogan's syndrome, psoriasis, Susac syndrome, Sjogren syndrome and ankylosing spondylitis.

Approximately all the cases with sudden SNHL are unilateral but there are evidence for bilateral involvement also. Mattox & Simmons (1997) reported less than 2% of patients have bilateral involvement and it is typically sequential whereas Fetterman et al. (1996) reported the prevalence of bilateral involvement was 1.7% to 3% and he reported bilateral simultaneous symmetrical hearing loss between both sides. The patients with sudden bilateral SNHL are more likely to have positive antinuclear antibodies, which indicating an association with immune-mediated aetiology (Fetterman et al., 1996). In the present review, the study done by Rossini et al (2016) reported 3 cases with sudden bilateral SNHL caused by systemic autoimmune diseases (SAD) where one had simultaneous involvement with Systemic Lupus Erythematosus (SLE) and two had contralateral posterior involvement 15 days after the first attack who had the underlying disease as Susac syndrome and other had Cogan's syndrome. Hence the prevalence of sudden bilateral SNHL in SAD is considerable and it is needed to be list out the various diseases which can cause a sudden bilateral SNHL.

The pathophysiology of sudden SNHL in vestibular schwannoma is not yet thoroughly understood. Putative factors include neuronal conduction block, vascular compression, rapid tumor growth and immune or toxic effects on inner ear (Saunders

et al., 1995). Although vestibular schwannoma normally develops very slowly, hemorrhage, necrosis, and cystic degeneration can rapidly increase tumour volume, resulting in auditory nerve compression with varying degrees of reversibility (Saunders et al., 1995). As a result of the diseases mentioned above, the rapid growth of initially tiny tumors in the inferior vestibular nerve may compress high- and middle-frequency fibers preferentially before affecting low-frequency fibers due to their position. Among the reviewed article, Hosakwa et al. (2018) reported that the mean hearing loss at 125 Hz, 250 Hz and 500 Hz was significantly milder in patients with sudden SNHL and vestibular schwannoma who had a trough-shaped audiogram than in those with idiopathic sudden SNHL hence suggest that A trough-shaped audiogram in patients with sudden SNHL especially with minimal to mild low-frequency hearing loss could be an indication of acoustic neuroma.

The role of imaging in the diagnosis of sudden SNHL is still not well defined because it does not seem to affect treatment much, but an extensive MRI study of the audiovestibular system, including nervous pathway and of whole-brain pre-and-post paramagnetic contrast administration, is recommended to eliminate the wide spectrum of abnormalities that cause sudden SNHL (Cadoni et al., 2006). Labelling sudden SNHL as idiopathic MRI is crucial to rule out vestibular schwannoma as it can cause 2% of sudden SNHL and accompanying partial recovery (Fetterman et al., 1996). Cho et al. reported one patient with vestibular schwannoma had complete recovery after treatment and suggested that MRI evaluation of patients with sudden SNHL for IAC tumours cannot be omitted, even in patients with complete recovery of hearing. Gupta et al. also suggest that MRI should be performed in all patients with sudden SNHL irrespective of hearing recovery.



Sudden SNHL was often treated with exploratory tympanotomy in the context of head trauma, barotrauma, chronic otitis media with cholesteatoma, and in individuals with congenital inner-ear abnormalities (Schreiber et al., 2010; Lu et al., 1989). In the present review, the study done by Haubner et al. (2012) suggests that a safe exploratory tympanotomy procedure may result in hearing improvement in patients with sudden SNHL due to perilymphatic fistula and an established treatment regimen as high dose steroid.

The current review aimed to update the various etiological factors of sudden SNHL and to document the epidemiology of those aetiology. The present study summarizes that the patients with sudden SNHL can have many underlying conditions that may lead to other complications in life. Hence, all the professionals who deal with sudden SNHL must perform appropriate diagnostic evaluation and proper referrals to help the patients get a formal treatment and right recommendation.

## Chapter 5

### SUMMARY AND CONCLUSIONS

The present review aimed to document the evidence for aetiology of sudden SNHL from literature published in the last 10 years. A totally internet-based search was carried out for the literature in the database of Google Scholar, Pubmed and J-GATE from December 2020 to July 2021. Articles regarding the topic of aetiology of sudden SNHL were searched in the mentioned database. The articles published from 2011 to 2021 July were considered to select for the review. The data extraction was performed by Rayyan intelligent, systematic review software developed by Qatar Computing Research Institute (QCRI). Based on the inclusion and exclusion criteria, 7 articles were selected for systematic review from 1,943 articles obtained after the initial search. The NIH study quality assessment tool was used for the quality analysis of finalized articles.

The summary of all 7 selected articles identified idiopathic as a major cause for sudden SNHL and other causes reported in the reviewed articles are as follows, inner ear or intra labyrinthine haemorrhage, acoustic trauma, head injury, vestibular schwannoma or acoustic neuroma, meningioma, labyrinthitis, structural deformity, perilymphatic fistula and systemic autoimmune diseases such as Lupus erythematosus, rheumatoid arthritis, scleroderma, vitiligo, Cogan's syndrome, psoriasis, Susac syndrome, Sjogren syndrome and ankylosing spondylitis. The updated articles in the current study may help the audiologist perform appropriate diagnostic test batteries to find the causal link between the various aetiological factors and sudden SNHL.

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