

Ministry of Defence

Synopsis of Causation

Sensorineural Hearing Loss

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Disclaimer

This synopsis has been completed by medical practitioners. It is based on a literature search at the standard of a textbook of medicine and generalist review articles. It is not intended to be a meta-analysis of the literature on the condition specified.

Every effort has been taken to ensure that the information contained in the synopsis is accurate and consistent with current knowledge and practice and to do this the synopsis has been subject to an external validation process by consultants in a relevant specialty nominated by the Royal Society of Medicine.

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1. Definition

- 1.1. Hearing loss may be conductive, sensorineural, or central in origin. **Conductive hearing loss** is caused by abnormalities of the external ear or of the [ossicles](#) in the middle ear. **Sensorineural hearing loss (SNHL)** is attributable to malfunction of the inner ear and/or auditory (eighth cranial) nerve. **Central auditory dysfunction** is caused by defects in the auditory [brainstem](#) or cerebral cortex. **Mixed hearing loss** involves a combination of conductive and sensorineural factors. This synopsis will focus on sensorineural hearing loss.
- 1.2. Hearing is measured by determining the quietest sound that a subject can hear across a range of frequencies. Hearing is considered normal when sounds across the normal conversational frequencies can be heard at 25 decibels (dB) or lower.
- 1.3. Levels of hearing loss may be classified as mild, moderate, severe, or profound although there is no universal agreement on precise definitions for these terms. The Royal National Institute for Deaf People (RNID) uses the following definitions:¹
 - **Mild deafness.** Threshold of quietest sounds that can be heard lies between 25 and 39 dB. People with mild hearing loss have some difficulty following speech, mainly in noisy situations
 - **Moderate deafness.** Threshold between 40 and 69 dB. People with moderate hearing loss have difficulty following speech without a hearing aid
 - **Severe deafness.** Threshold between 70 and 94 dB. People with severe hearing loss rely a lot on lipreading, even with a hearing aid. British Sign Language (BSL) might be their first or preferred language
 - **Profound deafness.** Threshold 95 dB or greater. People with profound hearing loss may communicate by lipreading and/or BSL. BSL may be their first or preferred language
- 1.4. Hearing loss is a common condition. In total, there are nearly 9 million people in the UK who are classified as either deaf or hard of hearing, of whom nearly 6½ million are aged over 60 years. Mild to moderate hearing loss affects around 8¼ million people, whilst just under ¾ million people have severe to profound hearing loss. 1.4 million people use a hearing aid regularly. There are an estimated 50,000 BSL users in the UK.
- 1.5. Adults are most commonly affected. Over 40% of people have some degree of hearing loss by the age of 50 years, and this figure rises to over 70% by the age of 70 years. 3.3% of people aged over 50 years have severe or profound hearing loss, rising to 7.6% for those aged over 70 years. At the other end of the age spectrum, 20,000 children in the UK aged 0 to 15 years are moderately to profoundly deaf. Congenital hearing loss is the most common birth defect.² In one study, the prevalence in the UK of bilateral SNHL of at least 40dB was around 1.2 children per 1,000 live births,³ whilst a later report indicated that the prevalence rises to 1.65 per 1,000 live births by the ages of 9 to 16 years.⁴
- 1.6. Hearing loss in childhood can be defined as **prelingual** or **postlingual**, i.e. according to whether the condition developed before or after the person has learnt to speak. Prelingual hearing loss can lead to particular difficulties with speech and language development.

- 1.7. The aetiology of SNHL is markedly heterogeneous, featuring both **congenital** and **acquired** causes, which are explored further in sections 3 and 4. Genetic and environmental factors act independently and in combination to produce hearing loss. Thus the susceptibility to hearing loss as a result of factors such as noise and presbycusis may be influenced by genetic predisposition. The precise correlation is yet to be elucidated.^{5,6}
- 1.8. It is important to appreciate the sensitivities attached to deafness. Many deaf people whose first or preferred language is BSL consider themselves as part of the Deaf community and hold a conviction that deafness is simply another way of life. The Deaf community does not think of deafness as limiting or as a medical condition. It should be noted that the term "hearing-impaired" is considered inappropriate by members of the Deaf community, who prefer the terms deaf and hard-of-hearing.
- 1.9. **Definition of some terms used in the measurement of sound:**
- Sounds vary according to their **frequency** (the number of times per second that the air vibrates from the acoustic energy), **amplitude** (volume or loudness), and **duration**. Noise may be intermittent or continuous, steady or impulsive
 - Frequency is measured in **Hertz (Hz)**, the **SI unit** of frequency in which one cycle per second equals 1 Hz. One **kilohertz (kHz)** is equal to 1000 cycles per second. Low frequency sounds are heard as rumbles or roars, high frequency sounds as screeches. The normal range of human hearing is about 20 Hz to 20,000 Hz
 - As acoustic energy increases, the amplitude of sound waves increase and the ear senses louder noises. Sound intensity is measured on a logarithmic scale in **decibels (dB)**. Every 3 dB increase represents a doubling of sound intensity. A whisper is about 20-30 dB, normal conversation 60 dB, a busy street 80 dB, lawn mower 90 dB, road drill 100 dB, chain saw 110 dB, rock concert 120 dB, jackhammer 130 dB, gunfire 140 dB, and a jet aircraft taking off at 25 metres is about 140 dB. Exposure to high noise levels can damage hearing (see section 4.1)
 - The human ear hears different sound frequencies with varying sensitivity and is most sensitive to sounds in the frequency range around 1 kHz to 4 kHz. For this reason, sound meters are usually fitted with a filter. The most widely used sound level filter is the "A" scale, which is used for measuring sounds that persist over a time period. The "A" scale is less sensitive to very high and very low frequencies, thus resembling the frequency response of the human ear. In contrast, the "C" scale gives near equal emphasis to sounds at most frequencies and is used to measure large amplitude sounds of extremely short duration such as a gunshot. There is also a rarely used B-weighting scale. When noise is weighted according to the "A" and "C" scales, values are written as "**dB(A)**" and "**dB(C)**" respectively
 - Measurements of noise in the workplace are often expressed as a **time-weighted average (TWA)**, which represents the average of the sampled sound taken over an eight-hour period. The TWA expresses the sound intensity that, experienced constantly over an eight-hour working day, would cause the same hearing damage as the variable noises to which the worker is actually exposed
 - **Hearing level** is defined for a specified frequency and testing system as the sound pressure level or vibratory force level of a pure tone relative to that of a reference

zero (as defined by an International or National Standard). It is the dial setting of an audiometer at which sound is heard if the instrument has been properly calibrated, expressed as dB HL

- **Hearing loss** is the amount by which a person's hearing threshold level deteriorates as a result of some adverse influence. A hearing loss at particular frequency is the number of decibels by which a tone must be amplified for the person to hear it

2. Clinical Features

- 2.1. SNHL may be unilateral or bilateral, and onset can be sudden or progressive. Different sound frequencies may be affected to varying degrees, with the result that perceived sounds often become both weak and distorted. Individuals may find that their hearing varies from day to day or from one situation to another. In adults, speech may deteriorate over time because high-pitched speech sounds cannot be heard. Hearing loss that is present at birth or arises in early childhood can affect social and educational development as well as the acquisition of spoken language.
- 2.2. Adults who develop SNHL typically describe difficulty keeping up with conversations in a group, comments from their family about the loudness of the television volume, and trouble using the telephone. They may have difficulty in distinguishing certain sounds, particularly 't', 'd' and 's', so that similar words become confused. Hearing difficulty, particularly speech discrimination, is often increased in the presence of background noise. High-pitched sounds such as bells may not be heard and, in severe cases, useful hearing may be lost completely.
- 2.3. **Tinnitus** is a common symptom occurring at some time in about 30% of the adult population and increasing in prevalence with age. A strong association has been reported between tinnitus and SNHL (including noise induced hearing loss). Tinnitus may also present as a feature of acoustic trauma, otosclerosis, and Ménière's disorder, as a side effect of many drugs, from a variety of other causes, or for no apparent reason (idiopathic tinnitus). It appears that tinnitus has little effect on the ability to hear in everyday life in the majority of patients; however tinnitus can be associated with varying degrees of hearing loss and with distortion of sound. It may be intermittent or improve over time. Tinnitus is subjective and there is no standard test to demonstrate its existence or to measure its disabling effect. Consequently, the severity of tinnitus can only be assessed by a detailed history using non-directive questioning. Relevant details include date of onset, how often it is present, whether it interferes with sleep or concentration, and whether the patient has sought medical advice.⁷
- 2.4. Some individuals with hearing loss also experience vestibular symptoms, dizziness or vertigo, or other problems with imbalance. Hearing loss can lead to depression, social isolation, and increased risk of accidents.
- 2.5. Most cases of **sudden SNHL** are unilateral and tinnitus is present in around 70-80% of patients. The peak incidence occurs between 30 and 60 years of age. People with unilateral hearing loss have difficulty in localising sounds and processing out background noises. Some cases of sudden sensorineural deafness may be associated with an underlying genetic disorder predisposing the individual to sudden, fluctuating or progressive loss of hearing; these conditions include branchio-oto-renal syndrome and Pendred's syndrome.
- 2.6. **Ménière's disorder** is a symptom complex, a triad or tetrad, associated with fluctuating, usually progressive SNHL with tinnitus, and episodic rotational vertigo; in about 70% of cases the fourth symptom of aural fullness is also present. The condition is usually unilateral but both ears are affected in around 15% of cases. Loud sounds may be uncomfortable and seem distorted in the affected ear. Initially, the hearing loss is in the low frequencies, but higher frequencies are affected as the disease progresses. The

onset is typically between the ages of 20-50 years and men and women are affected in roughly equal numbers. Symptom severity varies considerably between patients, although vertigo tends to be the most troublesome manifestation.

- 2.7. Some cases of SNHL are detected incidentally during routine tests on people who have no subjective complaint of hearing loss.
- 2.8. **Pure-tone audiometry** (PTA) is the standard investigation that is used to gather evidence regarding the type and extent of hearing loss. Average normal hearing (0 dB) is taken as the reference level on audiometers, a population based norm. Typically, air and bone conduction are tested for both ears at 250, 500, 1000, 2000, 4000, and 8000 Hz (may be supplemented with testing at 3000 and 6000 Hz), using varying intensities ranging from -10 dB to +120 dB. Hearing is considered within the normal range if tones across these frequencies can be heard at 25 dB or lower.
 - 2.8.1. Marked constitutional variation in hearing has been demonstrated in normal subjects. There is evidence of up to 30 dB variation at 2 kHz in young otologically normal adults matched for age and sex.⁷
 - 2.8.2. Ideally there should be a gap of at least 24-48 hours between the most recent noise exposure and the test. In practice, the gap is often reduced to a minimum of 12 hours.
 - 2.8.3. In addition to PTA, speech audiometry and other measures of hearing function may be undertaken. Speech audiometry involves the presentation of calibrated speech material to the patient via an audiometer, the patient's accuracy of responses being scored in a variety of ways depending on the actual speech material. It is usually possible to cross correlate the results of pure tone audiometry with speech testing results.
 - 2.8.4. Other tests of auditory function include testing for the presence of [stapedius reflexes](#) and for the presence of oto-acoustic emissions.
 - 2.8.5. Audiometricians are qualified, trained individuals who work to quality-assured standards. There is an expectation that the audiometrician will comment on whether the subject's audiometric responses were precise and repeatable, and whether the audiogram was consistent with informal observations.
 - 2.8.6. A **clinical hearing test** is a useful method for providing confirmation that an individual's hearing loss is reasonably consistent with PTA findings. For example, if the hearing distance for a conversational voice (CV) is 1 metre, the hearing loss should be in the region of 60 dB. If the hearing distance for a CV is 2 metres, the loss should be approximately 50 dB.
 - 2.8.7. More complex physiological tests may be required in some cases, including those where audiometric responses are not precise and repeatable or where there is substantial inconsistency between the results of PTA and the clinical hearing test. Available tests include cortical or brainstem evoked response audiometry, in which an [electroencephalogram](#) is used to detect brain wave response to sounds. In the medico-legal context, **cortical evoked response audiometry (cERA)** is used most frequently.

- 2.9. Imaging tests may be required in certain situations. For example, computed tomography (CT) of the temporal bones can be useful in detecting temporal bone anomalies and malformations of the inner ear e.g. enlargement of the vestibular aqueduct and developmental anomalies of the [cochlea](#). Further complementary information may be obtained by magnetic resonance imaging (MRI), indicated particularly for the diagnosis of a [vestibular schwannoma](#).
- 2.10. A careful family history can help to determine whether hearing loss is genetic. Molecular genetic tests are now available for an increasing number of the genes that have been linked to hearing loss.

3. Aetiology – Congenital and perinatal factors

3.1. Background and main features of congenital SNHL

3.1.1. Congenital hearing loss may be genetic, non-genetic, or of unknown causation.

- Around 50% of cases of congenital hearing loss are genetic, 25% non-genetic, and 25% of unknown causation
- Non-genetic hearing loss arises as a result of a variety of adverse events that can occur during intrauterine development. Examples of such events are included in section 3.4
- As knowledge of the human genome increases, it is becoming apparent that a significant and increasing proportion of [idiopathic](#) cases is likely to be attributable to genetic nonsyndromic hearing loss⁸

3.1.2. Genetic hearing loss is classified as **syndromic** or **nonsyndromic**.

- Around 30% of genetic hearing loss is syndromic and the remaining 70% is nonsyndromic
- In syndromic conditions, hearing loss is associated with abnormalities in other organ systems. Syndromic hearing loss can be conductive, sensorineural, or mixed
- In contrast, nonsyndromic hearing loss occurs in isolation. Nonsyndromic hearing loss is almost invariably sensorineural and is usually associated with a mutation in a single gene

3.1.3. The inheritance of genetic hearing loss (both syndromic and nonsyndromic) may be [autosomal dominant](#), [autosomal recessive](#), [sex-linked](#), or [mitochondrial](#). Within the prelingual nonsyndromic hearing loss group, inheritance is 75-80% autosomal recessive, 20-24% autosomal dominant, 1% sex-linked, and less than 1% due to mitochondrial inheritance.⁹

3.1.4. Several hundred genes are involved in the biology of hearing and many different mutations in this large number of genes can cause hearing loss. To complicate matters further, the clinical features may vary between different individuals with the same mutation. Thus age of onset, clinical severity and disease progression may not be predictable. Some genes may be associated with a recessive disorder in one individual and dominant transmission in another. Mutations in the same gene may cause syndromic hearing loss in one individual and nonsyndromic hearing loss in another. Conversely, matching clinical features may result from a variety of mutations in a particular gene or even from defects in different genes, e.g. [Usher syndrome](#), in which mutations in at least 10 different genes have been described.¹⁰

3.1.5. A positive family history can be instrumental in the diagnosis of hereditary hearing loss. However, a family history of hearing loss is often absent in autosomal recessive conditions. Here the abnormal gene may be passed through the generations from one unaffected carrier to another. Eventually, a situation will arise by chance where both parents carry the same abnormal gene with a resultant 25% possibility of producing a hearing affected child. In view of the

complex genetics involved, when parents who are both deaf reproduce, 90% of their offspring do not exhibit hearing loss.^{6,11}

- 3.2. **Syndromic hearing loss.** Over 400 genetic syndromes that involve hearing loss have been described. The most common autosomal dominant form is [Waardenburg syndrome](#). The most common autosomal recessive forms are [Pendred syndrome](#) and [Usher syndrome](#). Features of several of the syndromic forms of SNHL are summarised at Appendix A.
- 3.3. **Nonsyndromic hearing loss.** The various gene [loci](#) for nonsyndromic hearing loss are designated DFN (for DeaFNess). Loci for genes that are inherited in an [autosomal dominant](#) manner are referred to DFNA, those inherited in an [autosomal recessive](#) manner as DFNB, and those subject to [sex-linked](#) inheritance as DFN. A number is placed after the designation indicating the order in which the locus was mapped.
 - 3.3.1. More than 50 genes have been mapped for nonsyndromic hearing loss, and those that have been characterised at the molecular level encode proteins of diverse functions.¹² The most common cause of autosomal recessive nonsyndromic hearing loss, accounting for around 50% of such cases, is DFNB1. This disorder is caused by mutations in the *GJB2* gene, which encodes for the protein connexin 26 and, less commonly, the *GJB6* gene, which encodes connexin 30.
 - 3.3.2. Connexins in the inner ear form gap junctions between cells and are thought to help recirculate ions in the [cochlear endolymph](#), allowing potassium that enters hair cells during sound transduction to be recycled. The altered function of connexins in DFNB1 affects the process of potassium recirculation, causing cell death and permanent hearing loss. Prelingual hearing loss ensues although rarely the onset of hearing loss is delayed into early childhood. Molecular testing for the *GJB2* and *GJB6* genes is available.^{8,13}
 - 3.3.3. Although hereditary hearing loss usually is present at birth or develops in early childhood, some cases may only become manifest in adult life, e.g. DFNA9, which is caused by mutations in the *COCH* gene.
 - 3.3.4. Some [mitochondrial](#) DNA mutations, including 1555A>G and 7445A>G mutations, have been linked to sporadic and familial nonsyndromic, adult-onset SNHL. It is believed that individuals who carry the 1555A>G mutation are particularly sensitive to the ototoxic effects of aminoglycoside antibiotics (see section 4.7.1).⁶
- 3.4. **Other [congenital](#) and [perinatal](#) causes of deafness**
 - 3.4.1. **Prematurity and low birth weight** are significant factors in the aetiology of SNHL in children. The [odds ratio](#) for deafness rises with low birth weight; below 2,500 g it stands at 4.5, below 1,500 g it is 9.6.¹⁴ Admission to a neonatal intensive care unit for 48 hours or longer has been shown to carry a major risk of hearing impairment.¹⁵ The risk of SNHL has been linked to reduced oxygenation associated with asphyxia at birth, difficult delivery, persistent pulmonary hypertension associated with mechanical ventilation, and conditions requiring [extracorporeal membrane oxygenation](#).

- 3.4.2. **Chromosome abnormalities:** Down's syndrome (Trisomy-21) is commonly associated with hearing loss. In addition to conductive loss, SNHL can emerge in early adulthood. Progressive SNHL is common in Turner syndrome (females with missing X chromosome), affecting approximately 40% of patients by their mid-40s.¹⁶
- 3.4.3. Hearing loss due to congenital **cytomegalovirus (CMV)** infection can be present at birth or can appear later, usually during the first year of life. Epidemiological evidence suggests that around 12% of all congenital SNHL is caused by intrauterine CMV infection. CMV infection acquired postnatally is common but is not associated with hearing loss.^{11,17,18}
- 3.4.4. Other **prenatal infections** that may cause congenital SNHL include syphilis, toxoplasmosis, and rubella. Hearing loss develops in around one-third of cases of congenital syphilis, although it does not usually become evident until well into childhood or even into adulthood. Congenital rubella is now rare due to the introduction of an effective vaccination programme.
- 3.4.5. **Foetal alcohol syndrome (FAS)** is caused by the [teratogenic](#) effects of alcohol on the developing foetus and is characterised by facial abnormalities, growth retardation, cognitive impairment, and learning disabilities. The condition arises in infants born to mothers who have a history of heavy alcohol use during pregnancy. FAS has been linked to conductive, sensorineural, and central hearing defects, with limited data showing SNHL in 28% of infants born with the condition.¹⁹
- 3.4.6. Hearing loss is a feature of classic [kernicterus](#). However, evidence is also available to suggest that SNHL and central auditory dysfunction can be associated with elevated [bilirubin](#) levels in the newborn even in the absence of manifestations of kernicterus.²⁰ It is thought that the bilirubin pigment selectively damages the [brainstem](#) auditory nuclei and may also damage the auditory nerve.²¹
- 3.4.7. Auditory neuropathy or auditory dys-synchrony is a disorder of auditory function recognised over the last 5 to 10 years. It is a condition of failure of neural transmission of the auditory signal from the [cochlea](#) to the higher level auditory centres.

4. Aetiology - Occupational and other acquired causes

- 4.1. **Noise trauma** represents the most common preventable cause of SNHL. **Noise induced hearing loss (NIHL)** develops over a period of several years as a result of exposure to continuous or intermittent loud noise. This process is distinct from **acoustic trauma** in which a sudden but permanent change in hearing occurs as a result of a single exposure to a sudden burst of sound such as an explosive blast. This topic is covered in greater detail in the Synopsis “Blast Injury to the Ears”.
- 4.1.1. Limited exposure to loud noise can cause a **temporary threshold shift**, that is a measurable loss of hearing that is restored after the noise exposure has ceased, usually within 24 hours. The presence of temporary threshold shift is an indication that sustained exposure to the noise could cause permanent damage to hearing.
- 4.1.2. The onset of **persistent hearing loss** is usually gradual, arising as a consequence of prolonged exposure to loud noise. Damage from chronic exposure to high sound levels is cumulative and the rate and total degree of hearing loss are dose dependent. However, significant gaps remain in the understanding of the quantitative relationship between noise exposure (in terms of both intensity and duration) and resultant sensorineural hearing loss.⁷
- 4.1.3. NIHL is always sensorineural, caused by significant damage to the hair cells inside the [cochlea](#). Once this damage has occurred, the hair cells do not regenerate and permanent hearing loss ensues. To compound matters, at the same time as being noise-exposed, the individual is also ageing, and age-related loss will be superimposed on noise damage.⁷
- 4.1.4. Clinically, NIHL does not differ from SNHL due to a number of other causes. The condition is typically bilateral, although asymmetric sources of sound, such as sirens or gunshots from a shoulder-fired weapon, can produce asymmetric loss. NIHL typically produces a “notching” of the audiogram, which characteristically occurs initially at 4 kHz and subsequently progresses to involve the 3-6 kHz region with recovery at 8 kHz. However, in some cases the notch may initially develop at either 3 kHz or 6 kHz.
- 4.1.5. The audiometric pattern seen in NIHL contrasts with changes that occur as a result of ageing, where the highest frequencies are affected first. However, accurate diagnosis cannot be made on the basis of audiometric appearances alone. A typical notch at 4 kHz is not present in all cases of NIHL, whereas it may be seen in situations unrelated to NIHL, such as sudden viral hearing loss or ototoxic drug exposure. Genetic cases may be seen with a loss initially at 4 kHz. Furthermore, the prominence of a notch attributable to NIHL may reduce over a prolonged period of noise exposure as the loss from noise expands to affect other frequencies and changes due to ageing are superimposed.^{7,22}
- 4.1.6. The rate of hearing loss as a result of noise is greatest during the first 10-15 years of exposure. NIHL is considered to increase significantly with chronic exposures above 85 dBA for an 8-hour time-weighted average. In general, longstanding continuous noise exposure is more damaging than interrupted

exposure, which allows the ear to have a recovery period. However, short exposures to very high levels of noise may produce significant loss e.g. in firefighters and the construction industry. Individual susceptibility to the harmful effects of noise varies widely and it is thus not possible to predict the likelihood of progression or its rate. The biological basis for individual variability remains unclear. Studies of twins have demonstrated that noise sensitivity aggregates in families and probably has a genetic component.^{22,23}

- 4.1.7. Noise is a common cause of [tinnitus](#), but the symptom can arise from a variety of other causes. In addition to occupational noise exposure, tinnitus can be also occasioned by leisure noise and acoustic trauma. It is unlikely that tinnitus is attributable to noise in those cases in which it commences more than a year after noise exposure has ceased.⁷
- 4.1.8. Over 1 million employees in Great Britain are exposed to levels of **noise at work** that could damage hearing. Estimates derived from a large sample of UK working age adults indicate that nationally around 153,000 males and 26,000 females aged between 35 and 64 years suffer severe hearing difficulties as a result of occupational noise exposure. Corresponding numbers for persistent tinnitus are 266,000 males and 84,000 females.²⁴
- 4.1.9. Historically, the industries that were most closely linked with claims to the Industrial Injuries Disablement Benefit scheme for NIHL were shipbuilding, coal mining, metal manufacture and engineering. With changing patterns of employment, the major occupations that are giving rise to current claims are metal machining, other machine operators, mineral extraction, building trades, and assemblers.⁷
- 4.1.10. The Control of Noise at Work Regulations 2005 came into force on 6 April 2006 to replace the Noise at Work Regulations 1989. The main change was to reduce exposure action levels by 5 dB. Full compliance with the new regulations would eventually eliminate occupational NIHL and the HSE aims that by 2030 there should be no new cases of work-related NIHL.²⁵
- 4.1.11. The provisions of the new legislation place the following responsibilities on employers:²⁶
 - Assess risks and take action to reduce the noise exposure that produces those risks
 - Make hearing protection available on request at a daily or weekly personal noise exposure of 80 dBA and ensure so far as is reasonably practicable that hearing protection is worn at a daily or weekly personal noise exposure of 85 dBA
 - Ensure that the legal limits on noise exposure are not exceeded (daily or weekly exposure of 87 dBA or peak sound pressure of 140 dBC, taking account of any reduction in exposure provided by hearing protection)
 - Carry out health surveillance where there is a risk to health
- 4.1.12. A full consensus has yet to be reached regarding the magnitude of risk of NIHL associated with **recreational noise**, in particular, the use of personal listening devices in young adults. Exposure to hazardous levels of noise may commence during childhood and children's hearing is susceptible to such exposure, as

demonstrated by the detection in a US study of noise-induced threshold shifts in one or both ears of 12.5% of children aged 6 to 19 years.²⁷ Exposure to loud music is particularly common in adolescents and young adults. Sound levels between 120 dB and 140 dB have been recorded at rock concerts and levels above 95 dB have been found in bars on weekend nights. Transient threshold shifts of over 10 dB have been observed after listening to headphones for 3 hours at typical output levels.²⁸ A statistically significant increase of average hearing thresholds has been reported in young people using a personal cassette player for over 7 hours/week compared to those using one for 2-7 hours/week and compared to their matched controls. The same is true for subjects who go to rock concerts at least twice a month.²⁹

4.1.13. Individuals who engage in leisure activities that produce average sound levels greater than 90 dBA have been investigated and found to be significantly more likely to develop hearing loss than those who do not engage in noisy leisure pursuits. Thus, individuals who have engaged in woodworking are reported as being 30% more likely to have a hearing loss than those who have never followed this pursuit, with an increased risk of hearing loss calculated as 6% for each 5-year period of participation.³⁰

4.1.14. Permanent hearing damage can be caused immediately by a sudden, extremely loud, explosive noise, e.g. from explosions, airbag activation, gunfire or cartridge-operated machines. This topic is covered in more detail in the Synopsis “Blast Injury to the Ears”.

4.2. Exposure to a wide range of **chemicals** has been linked to SNHL. Inhaled chemicals and those absorbed through skin contact can reach the inner ear via the circulation.³¹ Exposure to a **combination of noise and ototoxic chemicals** occurs in a variety of occupations and research is ongoing to determine whether these factors may act synergistically to damage hearing. [Tinnitus](#), a sign of abnormal [cochlear](#) function, has also been associated with combined noise and chemical exposure. A particular concern is that noise exposure and toxic agents present in the workplace within current permitted levels could possibly pose an increased threat when encountered in combination.

4.2.1. Currently, ototoxic properties have been investigated for only a very small proportion of industrial chemicals. Experimental evidence has demonstrated that hearing may be affected by a variety of chemicals when present in sufficiently high concentrations. These substances include organic solvents (such as toluene, styrene, xylene, trichloroethylene, and ethyl benzene) asphyxiants (hydrogen cyanide and carbon monoxide) and metals (lead and mercury).³¹

4.2.2. The main focus of research has been on the manufacturing and construction sectors. Results have so far proved equivocal. For example, a review of evidence regarding styrene has found four occupational investigations that failed to detect any effect of styrene on hearing thresholds and three other investigations that claimed to demonstrate styrene-induced hearing loss in industrial populations, with synergism between styrene and noise. The reviewers noted shortcomings of experimental design and data analysis and concluded that hearing deficits due to occupational exposure to styrene at low concentrations have yet to be demonstrated by scientifically reliable argument.³² The NoiseChem project, approved and funded by the European Commission, is

intended to provide further data including an assessment of the dose/response relationships of a variety of chemicals, both alone and in the presence of noise.³³

- 4.3. Various researchers have investigated a possible association between NIHL and **vibration** (vibration white finger (VWF)/hand arm vibration syndrome (HAVS)). Although somewhat restricted in scope, the results have in general suggested that when exposed to noise, individuals with VWF/HAVS develop more hearing loss than those without VWF/HAVS. Limited evidence has also been published to suggest that the association between finger blanching and SNHL may extend to causes of blanching other than vibration induced white finger. The basis of these observations remains unclear.^{7,34}
- 4.4. **Presbycusis** is a gradual, progressive, bilateral hearing loss found in older people, usually commencing over the age of 65 years. The typical audiogram demonstrates a symmetric high-frequency hearing loss gradually progressing to middle and, eventually, to lower frequencies. Four pathological types of presbycusis have been described, these being sensory, neural, stria, and cochlear conductive.³⁵ The most consistent pathological finding is degeneration of sensory cells and nerve fibres at the base of the cochlea. Presbycusis is not attributable to a single cause but is understood to develop as a consequence of a combination of inherited and environmental factors. It remains unclear how much of the loss is due to physiological ageing, and how much from cumulative exposure to noise throughout the person's life. It is thought that mitochondrial mutations may contribute to presbycusis by compromising the availability of adenosine triphosphate (ATP), which provides the energy source required for maintenance of the ionic gradient within the cochlea.
- 4.5. **Trauma.** Hearing loss and vestibular disturbances are well-recognised complications of head injury. Symptoms are more common if a fracture of the [temporal bone](#) has occurred (especially a transverse fracture) than in fractures of other skull bones or in injuries that have produced brain concussion alone.³⁶ Even more specifically, SNHL has been found to be more prevalent in fractures involving the [petrous](#) portion of the temporal bone than in nonpetrous temporal bone fractures.³⁷
 - 4.5.1. Temporal bone fractures can cause conductive or sensorineural hearing loss. SNHL ensues when the fracture line involves the bony labyrinth (cochlea or vestibule).
 - 4.5.2. Conductive deafness caused by head injury may occur as a result of blood in the middle ear (haemotympanum), in which case the loss usually resolves within one to two months. However conductive deafness may also occur as a result of disruption of the [ossicular](#) chain, most commonly at the incudo-stapedial joint, or rarely as a result of massive ossicular disruption; in either case hearing loss will be permanent. SNHL that develops following head trauma may either improve or deteriorate within the first year of the injury, but thereafter the associated hearing loss can be considered to have stabilised.³⁸
 - 4.5.3. Trauma may lead to sudden SNHL in the absence of head injury (e.g. barotrauma due to SCUBA diving or diving into a swimming pool) in association with rupture of the [round or oval window](#) membranes and leakage of [perilymph](#).³⁹

- 4.6. A number of **infectious diseases** occurring in childhood or adult life may cause SNHL. These include the following:
- 4.6.1. Deafness occurs in 6% or more of childhood cases of **bacterial meningitis**. The inflammation may spread and destroy nerve cells associated with hearing. The organisms involved include *Neisseria meningitidis*, *Streptococcus pneumoniae* and *Haemophilus influenzae*, although the incidence of the last named has reduced following the introduction of immunisation.
 - 4.6.2. Permanent damage to the cochlea may be caused by childhood diseases that result in prolonged exposure to high fever e.g. **roseola infantum**, an infection caused by human herpesvirus type 6. SNHL has also been reported as a complication of infectious mononucleosis and measles.
 - 4.6.3. **Otitis media** typically causes a temporary conductive hearing loss, but chronic otitis media may occasionally cause permanent SNHL, either due to the disease process or, rarely, secondary to a surgical procedure.
 - 4.6.4. **Mumps** can give rise to profound SNHL, irrespective of whether or not the disease has been complicated by [meningoencephalitis](#). Transient high-frequency-range hearing loss has been reported in 4.4% of the cases of mumps in a military population. Permanent unilateral hearing loss occurs in 1 in 20,000 cases of mumps.⁴⁰
 - 4.6.5. SNHL may arise in AIDS and AIDS-related complex either as a direct consequence of **human immunodeficiency virus** (HIV) infection or as a result of opportunistic infection.⁴¹
 - 4.6.6. SNHL may develop gradually in untreated cases of **syphilis** acquired during adult life.
 - 4.6.7. Isolated reports have been published of SNHL following measles-mumps-rubella (MMR) vaccination but a definite link has not been established.⁴²
- 4.7. Various therapeutic **drugs** can damage hearing. In many cases, hearing will return to normal once medication is discontinued. The drugs that pose the greatest risk tend nowadays to be used in life-threatening situations only. As a general rule, high doses, prolonged treatment, and intravenous administration increase the risk of permanent damage.
- 4.7.1. Two major classes of drugs can cause permanent hearing loss by damaging the hair cells of the inner ear. These are aminoglycoside antibiotics (e.g. gentamicin, streptomycin, kanamycin, and neomycin) and platinum-based chemotherapeutic agents (e.g. cisplatin). Cell death is mediated by a complex pathway that involves the [mitochondria](#) (see also section 3.3.4).⁴³ It should be noted that the aminoglycosides may be either cochleotoxic (damaging to the hearing) or vestibulotoxic (damaging to the vestibular end organs).
 - 4.7.2. Other drugs that have been linked with ototoxicity include:
 - Salicylates (aspirin) when given in high doses. The mechanism appears to be multifactorial and individuals vary in susceptibility. Hearing loss is usually

accompanied by [tinnitus](#). Hearing is usually restored when medication is discontinued⁴⁴

- Several other nonsteroidal anti-inflammatory drugs (NSAIDs) including naproxen, piroxicam, and ketoralac have been linked to transient hearing loss and tinnitus. Very rare cases of sudden, permanent hearing loss have been reported with NSAID usage, usually in patients taking other ototoxic medications, with poor renal function, or with [autoimmune disease](#)^{45,46,47}
- Loop diuretics (e.g. furosemide), usually following intravenous administration
- Macrolide antibiotics (erythromycin), usually following intravenous administration
- Antimalarials (e.g. quinine, chloroquine). Hearing loss is often reversible on discontinuation of treatment
- Antifungal agents (e.g. amphotericin B) and antivirals (e.g. zidovudine, aciclovir)⁴¹
- Many chemotherapeutic agents

4.8. Miscellaneous causes

- 4.8.1. Patients who suffer from a variety of **autoimmune diseases** may rarely present with SNHL. In such cases, bilateral SNHL is rapidly progressive over a period of 3-4 months. Autoimmune processes may rarely target the [cochlea](#) specifically without causing symptoms in other organ systems. Conditions involved include Sjögren's syndrome, Wegener's granulomatosis, antiphospholipid syndrome, sarcoidosis, systemic lupus erythematosus, and Susac's syndrome. The underlying mechanism remains poorly understood but has been linked to immune-complex deposition in the cochlea.⁴⁸
- 4.8.2. **Otosclerosis**, which is characterised by abnormal bone growth, typically involves the [stapes](#). Resultant lack of mobility of the stapes causes a conductive hearing loss. However, involvement of the cochlea by otosclerosis may also develop, and this produces SNHL.
- 4.8.3. Conductive hearing loss, caused either by erosion of the [ossicles](#) or other pathological processes within the middle ear, is present in 90% of patients with [cholesteatoma](#).⁴⁹ In addition, longstanding cholesteatomas expand and may erode bone over the lateral [semicircular canal](#) or cochlea to create a labyrinthine [fistula](#). SNHL, sometimes profound, can occur in 3-22% of patients with a disease-induced canal fistula. Treatment for cholesteatoma is surgical.⁵⁰ SNHL may also occur in chronic suppurative otitis media even without a fistula, probably as a result of toxins leaching across the [round window](#) membrane.
- 4.8.4. The aetiology of **Ménière's disorder** remains uncertain but increased fluid volume ([endolymphatic](#) hydrops) within the inner ear has been identified in this condition.
- 4.8.5. **Iatrogenic** complications from ear surgery are rare. However, injuries that cause hearing loss may occur, e.g. suction in an open semicircular canal, which can cause SNHL, vertigo and disequilibrium.⁵⁰
- 4.8.6. There appears to be a weak association between **diabetes mellitus** and hearing loss.⁵¹ Of particular note, maternally inherited diabetes can arise in the presence

of mutations in [mitochondrial](#) DNA and this condition may be associated with additional features of mitochondrial disease, including bilateral SNHL.

- 4.8.7. Hearing loss is common in patients with **Paget's disease**. Fifty percent of those patients who have temporal bone involvement will present with hearing loss, usually mixed but occasionally purely conductive or sensorineural.⁵²
- 4.8.8. SNHL has been linked to **haematological disorders** involving elevated blood viscosity or increased rigidity of the red blood cells. Thus SNHL has been reported in association with conditions such as sickle cell disease, Waldenstrom's macroglobulinaemia, and various forms of leukaemia. In the last named, haemorrhage and/or leukaemic infiltration may be involved in the process.⁵³
- 4.8.9. **Tumours** may cause SNHL. The eighth cranial nerve is a relatively common site for a [schwannoma](#) (benign acoustic neuroma). Most cases occur sporadically and cause unilateral SNHL but neurofibromatosis type II, an autosomal dominant condition, is characterised by hearing loss caused by bilateral vestibular schwannomas. Very rarely, a primary malignant tumour, such as a squamous cell carcinoma, may cause SNHL by invading the inner ear. Secondary meningeal carcinomatosis may involve the eighth cranial nerve causing progressive bilateral SNHL.
- 4.8.10. **Radiotherapy** for head and neck cancer may produce hearing loss that is most appreciable in the higher frequencies. The effect appears to depend on the radiation dose received by the inner ear.⁵⁴
- 4.8.11. SNHL is a common finding in patients with **chronic renal failure**, including those treated with dialysis.
- 4.8.12. Limited, and often conflicting, evidence has been published with regard to the possibility that there may be an increased risk of developing SNHL in association with cardiovascular disease, hypertension, and raised serum lipid levels.
- 4.8.13. **Cigarette** smokers appear to have an increased risk of hearing loss. Smoking may, to a slight extent, increase the risk for those exposed to excessive noise. There also appears to be an increased risk for those without any history of occupational noise exposure.^{55,56}
- 4.9. The aetiology of **sudden hearing loss** is multifactorial. A definite cause, such as a viral infection, vascular disorder or [cochlear](#) membrane rupture can be identified in around 15% of cases only. Rarely, a vestibular [schwannoma](#) is implicated and sudden SNHL is also an uncommon symptom of multiple sclerosis.⁴⁹ No specific cause can be found in the remaining 85% of cases, which are thus termed [idiopathic](#), although an [autoimmune](#) basis may be suspected in some.
 - 4.9.1. Sudden idiopathic hearing loss appears to arise more frequently during crisis situations or long-term stressful life situations. In particular, patients who have suffered from relapsing sudden hearing loss have tended to describe phases of stress prior to the acute event. The psychophysiological basis for any possible link has yet to be elucidated.^{57,58}

5. Prognosis

- 5.1. Prelingual hearing loss can be identified through universal screening of neonates. Early identification and timely intervention is essential for optimal development of language and communication skills and for other cognitive stages of development, as much of learning is dependent on the attainment of language. Treatment options include amplification or cochlear implantation as appropriate. Most infants who are identified through screening have parents with normal hearing.⁵
- 5.2. Hearing loss prevention programmes form a vital component of the strategy for tackling exposure to occupational noise and chemical hazards. The Control of Noise at Work Regulations 2005, which were described in more detail at section 4.1.11, place a legal obligation on employers in the UK to assess and identify measures to eliminate or reduce risks from exposure to noise so as to protect employees' hearing. Strategies for the prevention of occupational NIHL include:
 - Engineering controls such as the design of less noisy equipment and reducing vibration (damping)
 - Administrative controls such as reducing time spent in noisy areas
 - Personal protective equipment (hearing defenders). The use of foam-insert earplugs decreases noise exposure by 30 dB
 - Training
 - Regular health surveillance incorporating periodic audiometry. Early detection of a relatively small threshold shift can allow hearing conservation measures to be reinforced before progression to significant hearing loss has ensued
- 5.3. As occupational noise exposure continues, so too does ageing. Precise details are yet to be established of how noise induced and age-related SNHL combine. Most evidence indicates that noise induced hearing loss neither improves nor deteriorates once exposure to noise at work is discontinued. However, an individual may still be exposed to non-occupational sources of noise and hearing will continue to worsen due to ageing. These confounding factors create difficulties in attempts to focus exclusively on the effects of occupationally related SNHL.⁷
- 5.4. Hearing loss associated with [autoimmune disease](#) may improve on treatment with steroids or methotrexate. Oral steroids are also used in the treatment of unilateral sudden SNHL, with early treatment being linked to a better outcome.⁴⁹ Biphosphonates and fluoride may be used to treat cochlear otosclerosis. Antioxidant drugs may be beneficial in preventing the progression of hearing loss in some cases caused by mitochondrial DNA mutation.⁶
- 5.5. Medical treatment for Ménière's disorder remains unsatisfactory; it includes a low-salt diet, diuretics, and drugs such as betahistine which are believed to improve the microcirculation within the inner ear. Labyrinthine sedatives should be used during the acute phase to control symptoms of vertigo. Hearing aids may be ineffective because patients suffer from poor speech discrimination, as well as diminished tolerance to amplified sound. Surgical intervention or chemical labyrinthectomy with intratympanic gentamicin are considered if medical management fails.⁴⁹ In the event of the development of profound deafness, cochlear implantation should be considered.

- 5.6. Hearing returns to normal, or near normal, without treatment in about 70% of cases of sudden [idiopathic](#) SNHL. However, the longer the hearing loss persists, the less likely it is to recover.
- 5.7. There is no specific medical or surgical treatment for NIHL or presbycusis. Consequently management focuses on functional improvement. Hearing aids may prove beneficial and both analogue and digital models are available. A hearing aid that is designed merely to amplify sound will fail to correct distortion, so that some difficulty in understanding speech will persist. Hearing therapists are accessible at some NHS clinics to provide advice on communication, the use of a hearing aid, and lipreading. Classes in lipreading are also available but the skill can prove difficult for older people to learn. Specially adapted equipment may be useful e.g. amplified telephone or textphone, vibrating pager, and a system of flashing lights linked to the doorbell. Loop systems, which help to reduce background noise, have been fitted in many public places.
- 5.8. A cochlear implant may be considered for both children and adults if hearing loss is bilateral and profound, and acoustic hearing aids prove ineffective. This electronic device is implanted under the skin behind the ear with the electrode array inserted into the [cochlea](#) in order to stimulate the auditory nerve directly. However, cochlear implantation is not an appropriate treatment in every situation.
- 5.9. Genetic counselling may be offered to patients with genetic disorders to help them to make informed personal and medical decisions. It should be noted that many people in the Deaf community are opposed to genetic testing for deafness as well as to the use of cochlear implants. Many view deafness as a distinguishing characteristic and not as a medical condition in need of a “cure”, whilst some parents do not consider it desirable to take steps to “prevent” deafness in their offspring.¹¹
- 5.10. There is no cure for most cases of [tinnitus](#) although a measure of relief can be obtained from a variety of approaches including hearing aids, maskers, antidepressants, counselling and relaxation techniques.

6. Summary

- 6.1. Hearing loss is common, with nearly 9 million people in the UK being classified as either deaf or hard of hearing, of whom nearly 6½ million are aged over 60. Congenital hearing loss is the most common birth defect.
- 6.2. SNHL may be unilateral or bilateral, and onset can be sudden or progressive. Sounds may become both weak and distorted. The hearing difficulty is often increased in the presence of background noise. Accompanying tinnitus and/or balance problems may be present, depending on the underlying cause of the hearing loss.
- 6.3. The basis of SNHL is multifactorial. Leading causes include genetic disorders, noise exposure, and presbycusis. Susceptibility to hearing loss as a result of factors such as noise and presbycusis can be influenced by genetic predisposition.
- 6.4. Noise trauma represents the most common preventable cause of SNHL. Noise induced hearing loss develops over a period of several years as a result of exposure to continuous or intermittent loud noise. Sources of noise may be occupational and/or recreational. The quantitative relationship between noise exposure and resultant sensorineural hearing loss is not yet fully understood.
- 6.5. Concerns have been raised that exposure to a combination of noise and ototoxic chemicals may act synergistically to damage hearing but results of research have so far proved equivocal.
- 6.6. Following head injury, SNHL is most likely to arise as a sequela of fractures that involve the petrous portion of the temporal bone
- 6.7. Hearing loss prevention programmes form a vital component of the strategy for tackling exposure to occupational noise and chemical hazards. Most evidence indicates that, once exposure to noise is discontinued, hearing loss does not progress beyond that which would be expected from the added effects of age-related loss. The management of established NIHL and presbycusis focuses on functional improvement, with the use of hearing aids, lipreading, and specially adapted equipment.

7. Related Synopses

Conductive Hearing Loss

Blast Injury to the Ears

Otitis Externa

Otosclerosis

Vertigo

8. Glossary

autoimmune disease	Illness which occurs when the tissues are attacked by the body's own immune system.
autosomal dominant	A gene that expresses its effects when one copy is present; therefore only one parent need have the characteristic in order to pass it to the offspring.
autosomal recessive	A gene that is required in two copies to be active, and therefore can be inherited only when both parents carry the gene.
bilirubin	A by-product produced by the liver from the breakdown of haemoglobin and haemoproteins. The causes of a raised level in the newborn include Rhesus incompatibility.
brainstem	The lowest part of the brain, which merges with the spinal cord and consists of the medulla oblongata, midbrain and pons. The origin of cranial nerve VIII (auditory nerve) is at the border of the pons.
cholesteatoma	A benign condition of unknown cause involving an expanding mass of cholesterol crystals and keratinised skin in the middle ear space.
cochlea	The part of the inner ear that is concerned with hearing. The cochlea is filled with fluid and lined with tiny hairs (cilia) that move when vibrated and cause a nerve impulse to be generated.
congenital	Pertaining to conditions present at birth.
electroencephalogram (EEG)	A diagnostic test that measures the electrical activity of the brain using highly sensitive recording equipment attached to the scalp by fine electrodes.
endolymph	Watery fluid contained in the membranous labyrinth of the inner ear (c.f. <i>perilymph</i>).
extracorporeal membrane oxygenation	Application of a life support system that circulates the blood through an oxygenating system. Used in neonates for the treatment of primary pulmonary hypertension, meconium aspiration syndrome, respiratory distress syndrome, group B streptococcal sepsis, and asphyxia.
fistula	An abnormal passage or communication.
iatrogenic	Induced inadvertently by medical treatment or procedures.
idiopathic	Of unknown causation.

kernicterus	Rare condition that develops in cases of severe neonatal jaundice due to brain damage caused by the deposition of bilirubin (<i>q.v.</i>).
locus (-i)	The location of a specific gene on a chromosome.
meningoencephalitis	Inflammation of the brain and surrounding membranes (meninges).
mitochondrial inheritance	Mitochondria are small organelles found in the cytoplasm of cells and responsible for energy production and cellular respiration. They are independent of the nucleus and contain their own DNA. Mitochondrial mutations are always maternally inherited as sperm do not contain mitochondria.
odds ratio	A statistical method of comparing whether the probability of a certain event is the same in two groups. An odds ratio of more than one implies that the event is more likely in the first group.
ossicle	A small bone, particularly used in reference to the 3 tiny bones of the middle ear. Hence: <i>ossicular</i> . The ossicles of the ear are called the stapes (stirrup), incus (anvil) and malleus (hammer).
perilymph	The fluid that surrounds the membranous labyrinth of the inner ear.
perinatal	Referring to the time just before, during and immediately after birth.
petrous portion	One part of the temporal bone (<i>q.v.</i>), consisting of hard bone and containing the inner ear mechanism and internal auditory canal.
round and oval windows	<i>Round window</i> : the opening in the wall of the middle ear leading to the cochlea. <i>Oval window</i> : the opening in the middle ear leading to the vestibule.
semicircular canals	Three membranous semicircular tubes contained in the inner ear, concerned with equilibrium and the interpretation of the body's position in space.
sex-linked inheritance	Inheritance that results from a gene located on a sex chromosome. The term is most commonly applied to X-linked recessive inheritance, which causes diseases that usually affect males. This occurs because they have only one X-chromosome, and so a single recessive gene on that X-chromosome will cause the disease.
SI unit	A unit of measure in the International System of Units.
stapedius reflex	An involuntary muscle contraction that occurs in the middle ear in response to high-intensity sound stimuli.

temporal bone	Bone situated at the side and base of the skull, which encases the inner ear.
teratogenic	Relating to substances or agents that can interfere with normal embryonic development
tinnitus	A sensation of ringing, whistling, buzzing, or roaring in the ears that occurs without external stimulus.
vestibular schwannoma	Also known as acoustic neuroma, acoustic neurinoma, or acoustic neurilemoma. A benign encapsulated tumour of the VIIIth cranial nerve composed of Schwann cells. These cells are responsible for providing the insulating myelin sheath for nerves.

Appendix A Examples of syndromic hearing loss ^{9,10,59}

Name	Mode of inheritance	Common features
Waardenburg syndrome	Autosomal dominant	SNHL, dystopia canthorum (lateral displacement of the inner canthus of both eyes), and pigmentary abnormalities of the hair (white forelock), skin and iris. Most common type of autosomal dominant syndromic hearing loss.
Branchiootorenal syndrome	Autosomal dominant	Conductive, sensorineural, or mixed hearing loss in association with branchial cleft cysts or fistulae, malformations of the external ear including preauricular pits, and renal anomalies.
Stickler syndrome	Autosomal dominant	Progressive SNHL, progressive myopia, cleft palate, abnormal epiphyseal development, and premature degenerative joint changes.
Pendred syndrome	Autosomal recessive	Severe-to-profound SNHL, developmental abnormalities of the cochlea, and goitre with or without hypothyroidism. The most common syndromal cause of deafness.
Usher syndrome	Autosomal recessive	SNHL, balance problems, and retinitis pigmentosa, Causes hearing loss, usually from birth, and gradual loss of sight, usually commencing in late childhood or adolescence. There are three clinical subtypes, designated types I, II, and III respectively. The most common cause of deaf-blindness in adults.
Jervell and Lange-Nielsen syndrome	Autosomal recessive	SNHL and prolongation of the <i>Q-T</i> interval on ECG, causing syncope and increased risk of sudden death.
Biotinidase deficiency	Autosomal recessive	SNHL, seizures, hypotonia, ataxia, visual problems, skin rash, and alopecia. Treated by the daily addition of biotin to the diet.
Refsum disease	Autosomal recessive	Progressive SNHL, visual problems, polyneuropathy, and cerebellar signs. Caused by faulty phytanic acid metabolism. Treated by dietary restriction and plasmapheresis.
Alport syndrome	Usually X-linked but autosomal recessive and autosomal dominant inheritance also described	Progressive SNHL of varying severity, progressive glomerulonephritis leading to end-stage renal disease, and variable ophthalmic findings.

Mohr-Tranebjaerg syndrome	X-linked	SNHL, dystonia, optic atrophy, fractures, and learning disability.
Norrie disease	X-linked	Progressive SNHL, ocular symptoms including retinal hyperplasia, and learning disability.
Kearns-Sayre syndrome	Mitochondrial	SNHL, ophthalmoplegia and retinal disease, myopathy, dystonia, heart block, endocrine abnormalities (e.g. diabetes), and short stature.
MELAS	Mitochondrial	Mitochondrial encephalomyopathy, lactic acidosis, stroke-like episodes, and SNHL.
MERRF	Mitochondrial	Mitochondrial encephalomyopathy, myoclonus epilepsy associated with ragged-red fibres, and SNHL.

Other inherited conditions that may feature SNHL among a range of symptoms include the mucopolysaccharidoses (Hurler, Hunter, Sanfilippo and Morquio syndromes), Friedreich's ataxia, Charcot-Marie-Tooth disease, osteopetrosis, and achondroplasia.

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