DEFINITION

1. The terms **"sensorineural deafness, perceptive deafness, nerve deafness"** are frequently used as synonyms for this condition.

2. However, the word **"deafness"**, which tends to imply total loss of hearing as **"blindness"** implies total loss of vision, should preferably not be used, the term **"hearing loss"** being a more accurate description, implying that the condition can be of varying degree.

3. **Sensorineural hearing loss** is usually a symptomatic manifestation of organic lesions of the inner ear, the auditory nerve and its central connections in the brain.

CLINICAL MANIFESTATIONS

4. Depending on the nature of the organic lesion concerned, the onset may be either sudden or insidious and one or both ears may be affected. Also depending upon the nature of the organic lesion, there may be other manifestations such as tinnitus and/or vertigo.

5. Some cases are detected incidentally during routine tests on people who have no subjective complaint of hearing loss.

6. Most patients complain in the initial stages of difficulty in distinguishing certain words in a sentence. Later, high-pitched sounds such as bells may not be heard and, in severe cases, useful hearing may be completely lost. The hearing difficulty is often increased in the presence of background noise.

AETIOLOGY

7. Sensorineural hearing loss may be either congenital or acquired.

CONGENITAL

8. Many forms of sensorineural hearing loss are genetically determined by dominant or recessive autosomal inheritance or by sex-linked recessive inheritance. Although usually evident in childhood, some cases may only become manifest as late as the fifth decade of life. Diagnosis of hereditary hearing loss depends on the finding of a positive family history.

9. Congenital sensorineural hearing loss may result from mishaps during intra-uterine life such as infection (especially rubella), toxic, metabolic and endocrine disorders affecting the mother, as well as anoxia associated with Rhesus incompatibility and difficult deliveries.

ACQUIRED

10. Sensorineural hearing loss may be -

    10.1. inflammatory, resulting from otitis media, typhoid fever, syphilis, mycoplasma, chlamydia, or viral infections such as mumps, measles, chicken pox and herpes zoster.

    10.2. vascular with end-artery obstruction, resulting from hypertension and/or atherosclerosis. In such a case, the hearing loss occurs suddenly and is unilateral.

    10.3. haematological, in association with such conditions as polycythaemia rubra vera, sickle-cell disease and leukaemia.

    10.4. associated with connective tissue disorders such as systemic lupus erythematosus, polyarteritis nodosa.

    10.5. associated with metabolic disorders such as diabetes mellitus, hyperlipidaemia, hypothyroidism.

    10.6. ototoxic due to various drugs, including certain antibiotics and diuretic agents, alcohol, tobacco, quinine and marijuana.

    10.7. associated with otosclerosis. Rarely otosclerosis may produce hearing loss which is entirely sensorineural.

    10.8. associated with tumours of the acoustic nerve and in the brain.

    10.9. traumatic.

    10.10. associated with advancing age. This type is known as **presbyacusis** and is evidenced by a typical audiogram showing a symmetrical high-frequency hearing loss gradually sloping downward with increasing frequency.

    10.11. **Non-organic**. This term is used when objective methods cannot detect any degree of hearing loss and no organic lesion is found.

    10.12. idiopathic.

11. The sensorineural hearing loss in those conditions listed at paragraphs 10.1 - 10.7 is an integral part of those conditions and the cause is thus that of the underlying condition.

12. **Traumatic sensorineural hearing loss** may result from -

    12.1. direct trauma to the head and involving the inner ear or the auditory nerve.

12.2. indirect trauma to the cochlear sensory hair cells and nerve fibres by exposure to sudden loud noise or continual or repetitive exposure to excessive noise as in industry or weapon firing. Such trauma typically produces impairment of hearing acuity in the 3-6 kHz band. The impairment is known as threshold shift. The threshold shift which occurs at the time of the exposure is usually temporary and, on removal from exposure, may improve with a possible residual permanent shift. The degree of impairment is dependent upon the degree of exposure and individual susceptibility. Continual or repetitive exposure increases the probability and severity of the permanent threshold shift. In the initial stages, whilst a threshold shift will be detectable on audiometry, it may not be sufficient to cause a subjective hearing loss. Any threshold shift will be increased in later years by the additive effect of presbyacusis but, once exposure to excessive noise has ceased, the hearing loss resulting from the noise exposure does not, of itself, worsen.

13. **Presbyacusis** is, as has already been stated, a feature of advancing age and results from degeneration of the cochlear cells and nerve fibres brought about by the ageing process. There is some evidence that a genetic component is involved since the condition is more marked in some families than others.

14. **Non-organic hearing loss** is the result of feigning in an endeavour to obtain compensation or of psychogenic causes.

15. When hearing loss is labelled **idiopathic**, that simply means that no obvious cause can be detected.

CONCLUSION

16. Sensorineural hearing loss is a symptomatic manifestation of damage to the inner ear, the auditory nerve and/or its central connections in the brain. There are many and varied causes which have been listed above.

REFERENCES

Baloh R W. The Special Senses. In: Wyngaarden J B, Smith L H and Bennett J C (Eds). Cecil Textbook of Medicine. Philadelphia. W B Saunders Company. 19th Ed. 1992. p2107-2108.

Booth J B. Sudden and fluctuant sensorineural hearing loss. In: (Ed) Booth John B. Scott-Brown's Otolaryngology. 5th Ed. 1987. Vol 3. Otology. London. Butterworths. p387-434.

Burns W. Noise and Man. 2nd Ed. 1973. London. John Murray. p110-114.

Byrne J E T and Kerr A G. Sensorineural hearing loss. In: (Ed) Booth John B. Scott-Brown's Otolaryngology. 5th Ed. 1987. Vol 3. Otology. London. Butterworths. p381-386.

March 1999