

ENTITLEMENT ELIGIBILITY GUIDELINES

OTOSCLEROSIS

MPC 00643
ICD-9 387

DEFINITION

Otosclerosis is a primary disorder of the bone of the otic capsule characterized by new bone formation, often involving the footplate of the stapes.

DIAGNOSTIC STANDARD

Diagnosis by a qualified medical practitioner, and results of audiometric investigation, are required.

ANATOMY AND PHYSIOLOGY

The ear has three small ear bones or ossicles, i.e. the footplate of the stapes (shaped like a stirrup), the malleus and the incus. Otosclerosis is an overgrowth of part of the temporal bone of the skull, just in front of the oval window which lies between the middle ear and the inner ear. The footplate of the stapes, together with the other ossicles, transmits sounds from the eardrum to the inner ear. The footplate bone initially becomes fixed by fibrous tissue and is subsequently replaced by sclerotic bone. Heredity is important in Otosclerosis. A positive family history has been reported in approximately 55% of persons with clinical manifestations of Otosclerosis.

CLINICAL FEATURES

The onset of Otosclerosis is generally during the second and third decades of life, and is determined by noting clinical Otosclerosis based on symptoms of Conductive Hearing Loss. Conductive Hearing Loss is caused by stapedial fixation and is present in approximately ten per cent of all cases. The amount of hearing loss directly relates to the degree of immobilization of the stapedial footplate, i.e. the more rigid the eardrum the greater the hearing loss. The hearing loss may be ignored in the early stages of Otosclerosis.

The disease may also invade the inner ear and cause a pure Sensorineural Hearing Loss, which is called Labyrinthine or Cochlear Otosclerosis.

PENSION CONSIDERATIONS

A. CAUSES AND/OR AGGRAVATION

THE TIMELINES CITED BELOW ARE NOT BINDING. EACH CASE SHOULD BE ADJUDICATED ON THE EVIDENCE PROVIDED AND ITS OWN MERITS.

1. Idiopathic
The cause of Otosclerosis may be unknown.
2. Heredity
Heredity is the only known cause, although the genetic inheritance pattern is not clearly established.
3. Inability to obtain appropriate clinical management

B. MEDICAL CONDITIONS WHICH ARE TO BE INCLUDED IN ENTITLEMENT/ASSESSMENT

- hearing loss

C. COMMON MEDICAL CONDITIONS WHICH MAY RESULT IN WHOLE OR IN PART FROM OTOSCLEROSIS AND/OR ITS TREATMENT

- tinnitus

REFERENCES FOR OTOSCLEROSIS

1. Australia. Department of Veterans Affairs: medical research in relation to the Statement of Principles Concerning Otosclerosis, which cites the following as references:
 - 1) Austin DF (1991) Otosclerosis in *Diseases of the Nose, Throat, Ear, Head, and Neck*. Ballenger JJ(Ed.) Lea and Febiger: Philadelphia. 14th Edition. pp 1159-1169.
 - 2) Brookes GB (1982) Vitamin D deficiency and otosclerosis. *Otolaryngol Head Neck Surg*. Vol. 93. pp. 313-321.
 - 3) Daniel HJ, Shambaugh GE and Fisch U (1973) Fluoride and clinical otosclerosis. *Arch-Otolaryngol*. Vol. 98. No. 5. pp. 327-329.
 - 4) Elbrond O, Jensen KJ (1979) Otosclerosis and pregnancy. *Otolaryngol*. Vol 4(4) pp. 259-266.
 - 5) Fechner RE (1990) Upper respiratory tract and ear in *Anderson's Pathology*, . Kissane JM (Ed). The C V Mosby Company: St.Louis. Vol 2. pp. 1077-1094.
 - 6) Goycoolea MV (1991) Otosclerosis in *Otolaryngology* Vol II, Paparella MM, Shumrick DA, Gluckman JL, Meyerhoff WL [Eds], WB Saunders Company: Philadelphia. Third Edition, pp. 1489-1512.
 - 7) Gristwood RE, Venable WN (1983) Pregnancy and otosclerosis. *Otolaryngol*. Vol. 8. pp. 205-210
 - 8) Harris JP and Keithley EM (1993) Inner Ear Inflammation and Round Window Otosclerosis. *The American Journal of Otology*. Vol. 14(2) pp. 109-112.
 - 9) Kristensen HK and Jorgensen MB (1967) Irradiation and Otosclerosis. *Acta oto-laryngologica*. 63 pp.114-120.
 - 10) McKenna MJ and Mills BG (1989) Immunohistochemical evidence of measles virus antigens in active otosclerosis. *Otolaryngol Head Neck Surg*. Vol. 101. pp. 415-421.
 - 11) Michaels L (1992) Ear, Nose, and Throat in *Oxford Textbook of Pathology*. McGee JO'D, Isaacson PG, Wright NA (Eds). Oxford University Press: Oxford. p. 1110.
 - 12) Nager GT (1988) Osteogenesis Imperfecta of the Temporal Bone and its Relation to Otosclerosis. *Ann Otol Laryngol*. Vol. 97. pp. 585-593.
 - 13) Nassif PS, Shelton C and House HP (1992) Otosclerosis. Treating progressive hearing loss in young adults. *Postgraduate Medicine*. Vol. 91(8) pp. 279-295.
 - 14) Soifer N, Weaver K, Endahl GL and Holdsworth CE (1970) Otosclerosis a review. *Acta Otolaryngol Suppl Stockh*. Vol. 269. pp. 1-25.
 - 15) Wiet RJ, Raslan W and Shambaugh GE (1986) Otosclerosis 1981 to

Entitlement Eligibility Guidelines - OTOSCLEROSIS

1985. Our Four-year Review and Current Perspective. *Am J Otolaryngol.* Vol 7(3) pp. 221-228.
- 16) Wiet RJ (1973) Patterns of ear disease in the southwestern American Indian. *Arch Otolaryngol.* Vol. 105(7) pp. 381-385.
- 17) Wiet RJ and Causse (1982) Otosclerosis -the last four years. *Am J Otol.* Vol. 3(3) pp. 249-255.
- 18) Wright I (1977) Avascular necrosis of bone and its relation to fixation of a small joint: the pathology and aetiology of "otosclerosis". *J Path.* Vol. 123(1) pp. 5-25.
2. Canada. Department of Veterans Affairs. Medical Guidelines on *Hearing Loss*.
3. Fauci, Anthony S. and Eugene Braunwald, et al, eds. *Harrison's Principles of Internal Medicine*, 14th ed. Montreal: McGraw-Hill, 1998.
4. Paparella, Michael and Donald A. Shumrick, et al, eds. *Otolaryngology Vol II Otology and Neuro-Otology*. 3rd ed. Chapter 45. Philadelphia: W. B. Saunders, 1991.