Entitlement Eligibility Guideline Otosclerosis

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ICD-11 code: AB33

VAC medical code: 00643 Otosclerosis

Definition

Otosclerosis is a bony overgrowth of the otic capsule, most commonly of the footplate of the stapes. As the overgrowth develops, the stapes can no longer function as a piston, but rather rocks back and forth and eventually becomes totally fixed. This dysfunction results in conductive hearing loss.

For Veterans Affairs Canada (VAC) purposes, vertigo, hearing loss (HL) and/or tinnitus may present as part of the symptom complex of a diagnosed medical condition, or they may present as a primary stand-alone diagnosed medical condition. In those presenting with symptoms of vertigo, HL, and/or tinnitus, but with a known diagnosed cause (e.g. Meniere's disease), these symptoms are included in entitlement and assessment of the medical condition. Prior to adjudicating the entitlement and assessment of vertigo, HL, and/or tinnitus, or a diagnosed medical condition that may cause these symptoms, a close review of previously entitled medical conditions with potentially overlapping symptoms is required.

Diagnostic standard

A diagnosis from an ear, nose and throat specialist ear, nose and throat (ENT) specialist/otolaryngologist is required.

The diagnosis is generally based on clinical findings and a formal audiogram. The audiogram shows conductive hearing loss, and may include a conductive notch at 2000 hertz (Hz).

A computed tomography (CT) scan may also help confirm the diagnosis, but a normal CT does not rule out the diagnosis.

Anatomy and physiology

The middle ear has three small ear bones or ossicles:

- the stapes, also known as stirrup
- the malleus, also known as hammer
- the incus, also known as anvil.

These bones transmit the vibrations produced by sound waves from the tympanic membrane to the oval window of the cochlea.

In otosclerosis, an overgrowth of bone occurs at the connection between the stapes and the oval window. This impedes the transmission of sound waves into the cochlea resulting in conductive hearing loss which increases over time.

Clinical features

The onset of otosclerosis is generally during the second and third decades of life. The etiology is not understood and may be multifactorial. The disease shows an inherited pattern of autosomal dominant inheritance with incomplete penetrance.

The amount of hearing loss directly relates to the degree of immobilization of the stapedial footplate, that is, the more rigid the stapes the greater the hearing loss. The hearing loss gets progressively worse.

Tinnitus is a common finding in otosclerosis, occurring in 50% of cases. Vertigo is present in about 10% of cases.

Most commonly, it affects both ears, with approximately 15% of cases affecting only one ear.

In about 10% of cases, the disease may progress and involve the inner ear and cause sensorineural hearing loss, which is also called retrofenestral, labyrinthine or cochlear otosclerosis.

There is an increased incidence (2:1) of clinical otosclerosis in females as compared to males.

Entitlement considerations

Section A: Causes and/or aggravation

For VAC entitlement purposes, the following <u>factors</u> are accepted to cause or aggravate the conditions included in the <u>Definition section</u> of this EEG, and may be considered along with the evidence to assist in establishing a relationship to service.

The factors have been determined based on a review of up-to-date scientific and medical literature, as well as evidence-based medical best practices. Factors other than those listed may be considered, however consultation with a disability consultant or medical advisor is recommended.

The timelines cited below are for guidance purposes. Each case should be adjudicated on the evidence provided and its own merits.

Factors

- 1. Having a **family history** of otosclerosis at the time of clinical onset or aggravation of otosclerosis.
- 2. Inability to obtain appropriate clinical management of otosclerosis.

Section B: Medical conditions which are to be included in entitlement/assessment

Section B provides a list of diagnosed medical conditions which are considered for VAC purposes to be included in the entitlement and assessment of otosclerosis.

- Hearing Loss
- Tinnitus
- Vertigo

Section C: Common medical conditions which may result, in whole or in part, from otosclerosis and/or its treatment

No consequential medical conditions were identified at the time of the publication of this EEG. If the merits of the case and medical evidence indicate that a possible consequential relationship may exist, consultation with a disability consultant or medical advisor is recommended.

Links

Related VAC guidance and policy:

- Hearing Loss Entitlement Eligibility Guidelines
- Tinnitus Entitlement Eligibility Guidelines
- Vertiginous Disorders Entitlement Eligibility Guidelines
- Pain and Suffering Compensation Policies
- Royal Canadian Mounted Police Disability Pension Claims Policies
- Dual Entitlement Disability Benefits Policies
- Establishing the Existence of a Disability Policies
- <u>Disability Benefits in Respect of Peacetime Military Service The Compensation Principle Policies</u>
- <u>Disability Benefits in Respect of Wartime and Special Duty Service The</u> Insurance Principle – Policies
- <u>Disability Resulting from a Non-Service Related Injury or Disease Policies</u>
- <u>Consequential Disability Policies</u>
- Benefit of Doubt Policies

References as of 22 January 2025

Australian Government, Repatriation Medical Authority. (1996). Statement of principles concerning acoustic neuroma, reasonable hypothesis, No 13 of 1996. SOPs - Repatriation Medical Authority

Australian Government, Repatriation Medical Authority. (1996). Statement of principles concerning acoustic neuroma, balance of probabilities, No 14 of 1996. SOPs - Repatriation Medical Authority

Australian Government, Repatriation Medical Authority. (2016). Statement of principles concerning acoustic neuroma, reasonable hypothesis, No 61 of 2016. SOPs - Repatriation Medical Authority

Australian Government, Repatriation Medical Authority. (2016). Statement of principles concerning acoustic neuroma, balance of probabilities, No 62 of 2016. SOPs - Repatriation Medical Authority

- Biswas, R., Genitsaridi, E., Trpchevska, N., Lugo, A., Schlee, W., Cederroth, C. R., Gallus, S., & Hall, D. A. (2022). Low evidence for tinnitus risk factors: A systematic review and Meta-analysis. *Journal of the Association for Research in Otolaryngology*, 24(1), 81–94. https://doi.org/10.1007/s10162-022-00874-y
- DynaMed (Update 2023 Feb. 15). Otosclerosis. Retrieved January 24, 2024 from https://www.dynamed.com/condition/otosclerosis
- Fauci, A. S., Braunwald, E., Isselbacher, K.J., Wilson, J.D., Martin, J.B., Kasper, D. Hauser, S.L., & Longo, D.L. (1998). *Harrison's principles of internal medicine*. (14th ed.).

 McGraw-Hill.
- Ferri, F. F. (2024). Ferri's Clinical Advisor Otosclerosis.

 https://www.clinicalkey.com/#!/content/book/3-s2.0-B9780323755764020044
- Hanna, B. (2023). Otosclerosis. *DynaMed*.

 https://www.dynamed.com/condition/otosclerosis#GUID-62EC1D4C-C7F2-4EA0-A524-D6D03A920827
- Insalaco, L. F. (2024 Jan.). Clinical Overview: Otosclerosis (Otospongiosis). Retrieved

 January 24 2024 from

 https://www.clinicalkey.com/#!/content/derived_clinical_overview/76-s2.0-

 B9780323755764006669#hl0000084
- Komune, N., Ohashi, M., Matsumoto, N., Kimitsuki, T., Komune, S., & Yanagi, Y. (2012).

 No evidence for an association between persistent measles virus infection and otosclerosis among patients with otosclerosis in Japan. *Journal of clinical microbiology*, 50(3), 626–632. https://doi.org/10.1128/JCM.06163-11

- Liu, X., & Yan, D. (2007). Ageing and hearing loss. *The Journal of Pathology*, 211(2), 188–197. https://doi.org/10.1002/path.2102
- McCormack, A., Edmondson-Jones, M., Somerset, S., & Hall, D. (2016). A systematic review of the reporting of tinnitus prevalence and severity. *Hearing Research*, 337, 70–79. https://doi.org/10.1016/j.heares.2016.05.009
- Paparella, M. M., Shumrick, D. A., Gluckman, J. L., & Meyerhoff, W. L. (1990). *Otolaryngology:*Otology and Neuro-otology v. 2 (3rd ed.). W. B. Saunders.
- Webber, P. C., Deischler, D.G., & Givens, J. (2024 Feb.) Up to Date: Etiology of Hearing
 Loss in Adults. Retrieved March 1, 2024
- World Health Organization. (2019). *International statistical classification of diseases*and related health problems (11th Revision). https://icd.who.int/
- Yong, J., & Wang, D.-Y. (2015). Impact of noise on hearing in the military. *Military Medical Research*, 2(1), 6. https://doi.org/10.1186/s40779-015-0034-5
- Zanon, A., Sorrentino, F., Franz, L., & Brotto, D. (2019). Gender-related hearing, balance and speech disorders: A review. *Hearing, Balance and Communication*, 17(3), 203–212. https://doi.org/10.1080/21695717.2019.1615812