

# ENTITLEMENT ELIGIBILITY GUIDELINES

## ACOUSTIC NEUROMA

**MPC**            00225  
**ICD-9**        225.1

### DEFINITION

Acoustic Neuroma is a sporadic, progressively enlarging benign tumour, that arises within the internal auditory canal from Schwann cells of the vestibular division of the eighth cranial nerve (acoustic nerve). It is also known as Acoustic Schwannoma, Acoustic Neurolemmoma, and Vestibular Schwannoma.

### DIAGNOSTIC STANDARD

Diagnosis by a qualified medical practitioner is required.

Investigations may include any of the following: skull x-ray, CT scan, MRI, lumbar puncture for CSF protein (which is always high - often over 200 mg/100 ml), and brain stem auditory evoked responses. Reports should accompany the application. Many years ago, the diagnosis of Acoustic Neuroma was rarely made in the absence of neurological symptoms.

### ANATOMY AND PHYSIOLOGY

Acoustic Neuroma is a benign slow growing tumour. It arises from the myelin-forming Schwann cells which form a sheath or coating for the acoustic nerve fibres. An Acoustic Neuroma will grow and expand into the cerebellopontine angle. A number of lesions of the cerebellopontine angle will present with unilateral otologic complaints and it will not always be possible to distinguish these preoperatively from Acoustic Neuromas.

## CLINICAL FEATURES

Early symptomatology may be subtle and easily escape detection. The growth rate is generally slow and meticulous. The history often reveals symptoms going back many years. Signs and symptoms develop primarily because of pressure effects that result from increasing tumour size.

Hearing Loss has been regarded as the hallmark of Acoustic Neuroma. The most common presentation is the development of Tinnitus associated with a unilateral, slowly progressive, high frequency Hearing Loss. The person may report increasing difficulty when conversations are directed to the affected ear or inability to use the telephone in the affected ear. However, any pattern of hearing loss may develop, including sudden hearing loss.

Routine audiometry may be normal or show a Hearing Loss of which the person is unaware.

Vertigo is an uncommon complaint. Minor disturbances of balance are common, with episodes of rotary vertigo being less common. These episodes may last several seconds or minutes to hours, with associated marked visceral autonomic symptoms.

Headache is, initially, usually occipital, but sometimes frontal. It may radiate from back to front through the mastoid region. In late stages it becomes general, and there may be attacks of severe occipital pain radiating down the spine with retraction of the head and neck, respiratory difficulties, and occasional loss of consciousness.

The most common neurologic deficits, outside of cranial nerve VIII, are changes in sensation within the distribution of cranial nerve V. Anesthesia of the posterior aspect of the external auditory canal is the earliest sign (*Histelberger's sign*), and, later, hyperesthesias and/or paresthesias of the face may occur. Deficits of the trigeminal nerve are more common than deficits of the facial nerve, but occasionally persons will manifest preoperative facial nerve weakness.

Early diagnosis is important as morbidity and mortality are directly proportional to tumour size. There is medical-scientific evidence that Acoustic Neuromas will continue to grow and may cause death if left untreated. They can now be detected in the early stages. Surgical removal is the norm, and in general the smaller the tumour the more successful the outcome.

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**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. Therapeutic radiation to head at least 10 years before clinical onset or aggravation

*Therapeutic radiation* means one or more treatments of radiation given with the aim of achieving palliation or cure with gamma rays, x-rays, alpha or beta particles.

It should be noted that the latency time for the development of Acoustic Neuroma, following therapeutic radiation to the head, has been recognized to be at least 10 years and up to 30 years.

2. Neurofibromatosis prior to clinical onset or aggravation

*Neurofibromatosis* (von Recklinghausen's disease) is an autosomal dominant genetic disorder with distinctive features. The features may be present at birth, as in café au lait lesions, or may develop decades later. One of these delayed features is Acoustic Neuroma.

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 ACOUSTIC NEUROMA AND/OR ITS TREATMENT**

- Tinnitus
- Vertigo
- Trigeminal nerve injury
- Facial nerve injury

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