

Clinical Effectiveness Guidelines

ACOUSTIC NEUROMA (Vestibular Schwannoma)



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GUIDELINE COMMISSIONERS

These Guidelines were commissioned by the Clinical Practice Advisory Group of the British Association of Otorhinolaryngologists – Head and Neck Surgeons (BAO-HNS).

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DISCLAIMER

The views represented in these guidelines are not necessarily the views of all members of the working party or their Associations. They are intended as a general guide to patient management and should not replace the clinical judgement professionals should exercise in managing individual patients.

MAIN RECOMMENDATIONS

1. Acoustic neuromas should be suspected in patients who present with unilateral or asymmetrical auditory symptoms (hearing loss or tinnitus). Magnetic resonance imaging represents the method of choice for identifying the minority of these patients who have an underlying acoustic neuroma.
2. Patients who are found to have an acoustic neuroma should be presented with the relevant management options available to them when they are initially diagnosed. Information about voluntary support agencies should be made available to patients.
3. The management options currently available include interval scanning, surgical removal or radiation treatment. The effect of each of these modalities is different and the selected strategy needs to be appropriate to the individual circumstances of the patient.
4. Management of these patients is best undertaken by dedicated multidisciplinary teams working in specialist centres.
5. There is need for prospective longitudinal outcomes research to compare the relative merits of the various management modalities, especially in those patients with small neuromas or where no tumour growth has been demonstrated.
6. A national audit of acoustic neuroma is needed to analyse outcomes across centres to ensure a uniformly acceptable standard of care is available to patients nationally. Validated measures that assess all aspects of outcomes (including quality of life issues) need to be developed and implemented in clinical practice.

1. OBJECTIVES

The purpose of these guidelines is as follows:

- 1.1 To assist professionals and patients about decisions relating to the diagnosis and management of acoustic neuromas based on the best available evidence. These guidelines may also be of value to health-care commissioners in their purchasing decisions.
- 1.2 To facilitate the early detection of acoustic neuromas.
- 1.3 To improve the quality and consistency of patient care and outcome across Great Britain and Ireland.

Note: The Guidelines do not cover the care of patients with Neurofibromatosis II or those patients with non-acoustic neuromas.

2. INTRODUCTION

- 2.1 Acoustic neuromas represent about 6% of all intracranial tumours. They are benign, slow-growing tumours, which arise from cells in the sheaths that surround the hearing and balance nerves. The neuromas usually manifest themselves as one-sided hearing impairment, which may go ignored by the patient or be dismissed by the doctor. Continued growth of these neuromas ultimately results in compression on the brainstem and a rise in pressure within the brain (raised intracranial pressure). Early diagnosis offers patients a range of management options and may significantly reduce morbidity.
- 2.2 Pathologically, acoustic neuromas are vestibular schwannomas. Because the term 'acoustic neuroma' is in such common usage, this term has been maintained throughout the text.

3. MOST FREQUENT PRESENTING SYMPTOMS

- 3.1 About 90% of patients present with gradual progressive hearing loss in one ear. However, about 5% will present with a sudden hearing loss. Many (about 70%) experience tinnitus (noise) in one ear. However, about 3 % of patients with acoustic neuromas will have normal hearing at presentation. Patients, especially when asked, will often admit to experiencing balance disturbance, which may be quite mild.
- 3.2 Less common presentations include facial numbness or pain from involvement of the trigeminal nerve, inco-ordination from cerebellar compression or earache. Facial weakness is very uncommon despite the fact that the neuroma is pressing on and causing damage to the facial nerve. Large or medially-placed neuromas may have symptoms of raised intracranial pressure such as headaches, visual disturbance or a diminution in the level of responsiveness.

- 3.3 The advent of Magnetic Resonance Imaging (MRI) has resulted in the diagnosis of symptomless acoustic neuromas in patients who were scanned for other unrelated conditions (Selesnick et al. 1993).

4. EPIDEMIOLOGY OF ACOUSTIC NEUROMA

- 4.1 The incidence of acoustic neuromas – the number of newly diagnosed cases per year – is around 13 cases/million/year (Moffat et al 1989, Davis 1995). Tos et al (1999) have shown that incidence figures in Denmark have increased over the last 20 years from 7.8 to 12.4 cases/million/year. This is probably a reflection of better diagnostic methods rather than a true increase in neuroma incidence.
- 4.2 These incidence figures are based on patients who have received a diagnosis whereas estimates of prevalence – the number of cases in the population – have so far been estimated only on the basis of large, unselected autopsy or radiology studies.
- 4.3 An autopsy study by Leonard and Talbot (1970), suggested a prevalence of 0.8 % or 8,000 per 1,000,000. If one accepts the clinical incidence of acoustic neuromas as being between 13 per 1,000,000, it is reasonable to assume that many neuromas escape diagnosis.
- 4.4 A recent radiological study found 7 unsuspected acoustic neuromas per 10,000 brain MRI studies – equivalent to 700 cases per 1,000,000 population (Anderson et al 2000).
- 4.5 The number of patients with asymmetrical hearing symptoms, who attend ENT clinics and are eventually diagnosed with acoustic neuroma, ranges from 3-7.5% (Ferguson et al 1996; Hollingworth et al 1998).
- 4.6 Within a District General Hospital ENT clinic it has been shown that 19.7% of new patients attending the ENT outpatient department are potential candidates for screening for acoustic neuromas (Harcourt et al 1999).
- 4.7 The National Study of Hearing showed that 2.9% of the population has an asymmetry greater or equal to 15 dB across 0.5 – 4 kHz. For the high frequencies 4, 6, and 8 kHz, this prevalence increases to 10.4% (Davis et al 2000). Where the better ear has hearing thresholds better than 25 dB, the prevalence values are 5.2% and 10.9% respectively.
- 4.8 These data highlight the need for effective screening of patients with asymmetrical hearing symptoms, as only a minority will prove to have an acoustic neuroma.

5. WHO SHOULD BE INVESTIGATED?

- 5.1 The diagnosis of acoustic neuroma should be considered in any patient complaining of unilateral or asymmetrical auditory symptoms (either hearing loss or tinnitus), whether of progressive or sudden onset in whom there is no other obvious cause. The diagnosis should also be considered in patients with impaired facial sensation and in certain patients with imbalance, which cannot otherwise be explained.

- 5.2 Age alone should not dictate whether or not a patient with appropriate symptoms is investigated. Knowledge of the diagnosis, even if no treatment is offered, may help in patient management.

6. WHAT INVESTIGATIONS ARE APPROPRIATE?

6.1 Audiological

6.1.1 Pure Tone Audiometry

All patients with unilateral or asymmetrical hearing symptoms should have a pure tone audiogram, appropriately masked as necessary. If previous audiometry is available, any change in threshold should be noted. Most patients have a high frequency sensorineural loss at presentation but any pattern of hearing loss (or normal hearing) may be encountered.

6.1.2 Speech Discrimination Testing

In the past, much emphasis was placed on speech audiometry for ‘site of lesion’ testing. However, it has largely been supplanted in this role. The test is still of great value to assess the usefulness of hearing in the neuroma ear, especially when hearing conservation surgery is being considered.

6.1.3 Electric Response Audiometry

Auditory brainstem responses (ABR) have been described as having high sensitivity and specificity (over 90%) in the diagnosis of acoustic neuromas (Hood 1998). However, careful determination of the false negative rate of ABR, indicates that especially for small neuromas it is unacceptably high and approaches 17% (Wilson 1997). Its usefulness is further limited by the fact that in cases of severe to profound hearing loss, no ABR response can be elicited due to the extent of the hearing loss. An absent ABR in the presence of milder losses, however, is suggestive of retro-cochlear pathology. There has been recent interest in the use of otoacoustic emissions to help assess the status of the cochlea in patients with acoustic neuromas.

- 6.1.4 ABR represents the non-imaging test of choice in the small number of patients in whom MRI may be contraindicated or not tolerated. The test is non-invasive, is well tolerated by patients, but requires sophisticated equipment and a high degree of expertise, especially in the interpretation of test results. Open MRI may also be an option for such patients (but the field strength of open magnets is significantly reduced and may be inadequate for screening purposes).

6.2 Diagnostic Imaging

6.2.1 Computed Tomography (CT)

CT scanning is performed using thin sections (2–3 mm) after intravenous injection of an iodine-containing contrast medium and filmed at soft tissue and bone window settings. It has the advantage of being widely available, cheaper than magnetic resonance imaging (MRI) and it shows bone erosion of the internal auditory canal (IAC) to best advantage. Some patients who cannot be examined by MRI (claustrophobia, cardiac pacemaker etc.) may be scanned by CT.

- 6.2.2 CT provides excellent visualisation of moderate or large (>15 mm) neuromas in the cerebello-pontine angle cistern with a sensitivity of detection approaching 100%. However, the sensitivity falls if the neuroma is smaller than 10 mm and CT is unable to detect those neuromas entirely confined to the IAC (intracanalicular neuromas).
- 6.2.3 In the majority of cases it is highly desirable to detect acoustic neuromas when they are small. This lack of sensitivity and the modest dose of ionising radiation imparted to the patient make CT an inappropriate method of screening large numbers of patients. However, depending on local availability and cost of MRI, CT may retain a role in scanning elderly patients where the aim is limited to detecting larger space-occupying lesions.
- 6.2.4 *Magnetic Resonance Imaging (MRI)*
MRI is the most accurate diagnostic test for identifying acoustic neuromas (Curtin 1997). It also has the advantages of multiplanar imaging, of enabling an assessment of the labyrinth, and it does not involve ionising radiation. It has largely supplanted CT in the investigation of patients with unilateral or asymmetrical audiovestibular symptoms. The two major approaches for screening patients in order to detect acoustic neuromas with MRI use T2-weighted images or contrast enhanced T1-weighted images.
- 6.2.5 One strategy is to acquire T2-weighted images only. The spatial resolution of the images must be sufficient to reliably demonstrate the cisternal and intracanalicular portions of the VII and VIII nerves, such that for small neuromas it is possible to identify which branch of the vestibulocochlear nerve (or rarely, the facial nerve) is involved. T2-weighted images also enable identification of whether there is residual CSF between the neuroma and the fundus of the IAC, which may influence surgical planning. Vascular compressive lesions, peritumoral oedema and associated arachnoid cysts are also revealed. Cochlear anomalies and large vestibular aqueduct syndrome, which may account for as many as 10% of abnormalities associated with sensorineural hearing loss, are easily identified (Daniels et al 2000). Although some intralabyrinthine neuromas can be visualised, very small neuromas may evade detection.
- 6.2.6 If T2-weighted images are to be performed, every care should be taken to optimise scan parameters. Employing a standard 2D FSE sequence with a slice width of 3 mm slice runs the dual risk of incurring a high false negative rate and having a high rate of equivocal scans requiring additional T1-weighted enhanced images (Zealley et al 2000). Scanner capabilities vary so much that it is impractical to specify parameters. However, close attention needs to be paid to slice thickness (maximum 2 mm), slice interleaving, matrix size (e.g. 512 x 512), number of acquisitions (e.g. 4-6), and field of view (maximum 20 x 20 cm) along with the possibility of using dual phased array receiver coils over both temporal bones. 2D images should be acquired in axial and coronal planes.
- 6.2.7 Scans should be interpreted by appropriately experienced neuroradiologists or ENT radiologists, and patients recalled for contrast enhanced imaging in cases of radiological uncertainty.
- 6.2.8 However, if available, a single acquisition using a T2-weighted 3D FSE or T2*-weighted CISS technique offers considerable advantages in spatial resolution (slice thickness <1 mm) with potential for image reconstruction and slice overlap to show the nerves and labyrinthine structures in greater detail. High resolution 2D or 3D T2-weighted techniques,

in conjunction with appropriate clinical pre-screening and referral, can provide a very sensitive method of evaluating unilateral sensorineural hearing loss compared to contrast-enhanced T1-weighted images (Daniels et al 1998 & 2000). It is also advisable to perform a T2-weighted FSE (or similar) sequence through the brain especially if the brainstem has not been well visualised by the focussed high resolution images.

- 6.2.9 As only a limited number of sequences are employed, examination times are short. Medical staff need not supervise the scans and if suitable patients are “batched”, large numbers can be scanned in a given MRI session. However, this approach may not be suitable for older generation or lower field (< 1T) scanners where spatial resolution is insufficient to clearly define the individual nerves. It may prove necessary in a small percentage of patients to obtain additional contrast-enhanced T1-weighted images patients with equivocal findings on T2-weighted images or where patient movement leads to an inability to resolve the individual components of the nerve complex. For scans that are medically unsupervised, this requires patients to be recalled. It should be recognised that radiologists reporting these scans will have varying degrees of training and familiarity with imaging this region, and some may have a preference for and greater confidence with reporting contrast-enhanced T1-weighted images.
- 6.2.10 The other approach is to only acquire T1-weighted images. Injection of a gadolinium-based contrast medium increases the sensitivity of detecting small neuromas and intralabyrinthine lesions, non-enhancing schwannomas being extremely rare. It is considered by most to be the gold standard for detecting acoustic neuromas. Inflammatory disorders of the VIII nerves and labyrinths may also be revealed. The anatomical detail of the nerves and fluid containing structures however is much poorer than on T2-weighted images.
- 6.2.11 T1-weighted images are acquired in the axial plane, and often the coronal plane, using 2-3 mm sections. Preliminary unenhanced T1-weighted images may help detect rare lesions such as lipoma or labyrinthine haemorrhage but many centres in the UK routinely acquire T1-weighted images only after intravenous injection of contrast medium. The gadolinium-containing contrast media are well tolerated with an extremely low incidence of side effects.
- 6.2.12 Injecting a contrast agent increases overall examination time and requires medical staff to inject or be in close proximity if it is to be administered by radiographic staff. This may preclude scheduling of scans outside normal working hours. The use of contrast medium increases the cost of the examination although it is not necessary to inject as much as 0.1 mmol/kg. Five-ten ml of contrast agent is sufficient, limiting the additional expense to £20-30.
- 6.2.13 In centres with a substantial otolaryngology service, MRI requests for patients with suspected acoustic neuromas might comprise 10% or more of all MRI examinations. Whilst T1-weighted and T2-weighted images can provide complimentary information, the routine use of both techniques may be difficult to justify for screening purposes in the face of limited resources. Selecting the appropriate screening protocol will depend upon local circumstances (e.g. type of scanner, MRI session availability, intravenous (I.V). injection protocols, radiologist’s skills and preferences) and upon balancing the small risk of missing a very small intracanalicular or intralabyrinthine neuroma against cost and increasingly long waiting times for investigation and diagnosis. There is an increasing

consensus that T2-weighted imaging, if of a sufficiently high quality, can be employed as a highly accurate and cost effective screening test where the overall demand for MRI services is very high (Daniels et al 2000).

7. MANAGEMENT OPTIONS

- 7.1 There are three management options for acoustic neuroma patients:
 - a. Interval scanning,
 - b. Surgical removal of the neuroma or
 - c. Stereotactic radiosurgery / stereotactic fractionated radiotherapy
- 7.2 The major determinants of which treatment is adopted are: neuroma size, age, health-status, the desire to attempt hearing preservation, the state of hearing in the opposite ear, and the preference of the patient after due consideration of the advantages and risks of each option.
- 7.3 Patients should be presented with the full range of management options available to them when they present.
- 7.4.1 **No intervention with interval scanning**
- 7.4.2 This strategy may be considered, at least in the short term, for patients with a small neuroma and good hearing. Older patients and individuals in poor health may be managed in this way, certainly initially, although it is by no means certain that neuromas will grow less aggressively in old people.
- 7.4.3 For intracanalicular neuromas, an observation period between scans of approximately 1 year may be appropriate as there is evidence that some of these neuromas cease to grow, at least in the short term. (O'Reilly et al 2000).
- 7.4.4 Spontaneous involution of acoustic neuromas has been reported in up to 13% of patients (Luetje 2000).
- 7.4.5 In one study, two-thirds of neuromas did not grow over a mean follow-up period of 35 months and neuroma involution was observed in 12% of cases. Growth in the first year following diagnosis was predictive of later neuroma enlargement. The authors recommended that in the absence of documented growth, watchful waiting represent the best management option. (Tschudi et al 2000).
- 7.4.6 Evidence suggests that, using the middle fossa approach, a hearing conservation rate of 69% can be achieved in small neuromas with good pre-operative hearing (Weber et al 1996). Allowing such neuromas to grow, especially in young patients, could compromise the ability to preserve hearing.
- 7.4.7 All patients being managed conservatively should be reviewed by annual imaging, to look for neuroma growth. Neuromas demonstrating growth (as demonstrated on serial scans by an increase in cross-sectional diameter or by an increase in neuroma volume) should then

be considered for either surgery or radiotherapy. However, there is no agreed, validated measure of neuroma size across centres.

7.5 Surgical Removal

- 7.5.1 Surgical removal is the treatment offered to the majority of patients with acoustic neuromas. Two surgical approaches predominate in the UK: the retrosigmoid (RS) and translabyrinthine (TL). Other less frequently employed operations are the Middle Fossa (MF), the Extended Middle Fossa (EMF) and Transotic approaches.
- 7.5.2 The RS, MF and EMF approaches provide the chance of preserving residual hearing in a subset of patients with good pre-operative hearing and suitably located neuromas – the chances of succeeding in this particular subgroup of patients being no greater than 50%. Hearing conservation should also be considered in patients who have poor hearing in the contralateral ear. There are no agreed audiological criteria for hearing conservation. The RS or MF approach is therefore employed for patients with useful residual hearing and a reasonable expectation of hearing preservation. The TL is generally employed for patients with poor hearing and /or larger neuromas where hearing preservation is not achievable.
- 7.5.3 Complete neuroma removal is achievable in over 95% of cases (Hardy et al 1989).
- 7.5.4 Operative mortality in experienced centres is less than 1% with the risk being slightly greater with larger neuromas (Ramsden 1995). A possible risk of epilepsy attends middle fossa approaches.
- 7.5.5 Permanent facial paralysis, either partial or complete, remains the greatest single source of disability following neuroma removal; those patients with large neuromas are at greatest risk of this complication (Cerullo et al 1993; Lalwani et al 1994; Nutik et al 1994). Other factors, such as the degree of adherence and intermingling between tumour and nerve, make precise pre-operative prediction difficult in individual patients. The most commonly used clinical grading system for facial paralysis, the House Brackmann Scale (House et al 1985), should be used in reporting of results.
- 7.5.6 A learning curve has been described in acoustic neuroma surgery with a plateau being reached between 20 and 60 cases (Welling et al 1999, Buchman et al 1996). A surgeon commencing acoustic neuroma surgery should be appropriately trained, preferably having carried out an appropriate number of operations under supervision prior to establishing an independent practice. Surgery is often undertaken as a collaborative exercise between the neuro-otologist and the neurosurgeon, especially for large neuromas.
- 7.5.7 The use of facial nerve monitoring has improved the outcome of patients undergoing acoustic neuroma surgery and its use is considered mandatory for any acoustic neuroma operation (Kartush 1998). Monitoring cochlear function may also be useful in hearing conservation surgery.

7.6 Stereotactic radiosurgery (SR) and fractionated stereotactic radiotherapy(FSR)

- 7.6.1 Stereotactic radiosurgery (SR) was defined by its pioneers (Leskell and Larsson) as the application of single fraction ionising radiation to a stereotactically defined volume of tissue, irrelevant of its histological composition.

- 7.6.2 More recently, techniques have been refined to enhance treatment of the neuroma and reduce the risk of radiation damage to surrounding structures. This can be achieved by reducing the dosage of radiation, using a stereotactic technique to treat irregular lesions in their three dimensions, and giving multiple small doses – fractionated stereotactic radiotherapy (FSR). Image fusion to optimise shaping and planning should be a standard of care if radiotherapeutic approaches are used. Small and medium sized neuromas up to 3.0cm in diameter are considered as being potential candidates for SR or FSR treatment (Forster et al 1996).
- 7.6.3 The source of radiation in SR and FSR is either gamma ray photons from multiple high activity Cobalt-60 sources or a linear accelerator (LINAC) which uses X-ray photons derived from high-energy electrons. Although both sources can be used for SR and FSR, in practice the Cobalt source is almost exclusively used to deliver SR.
- 7.6.4 SR and FSR do not remove neuromas but are generally proposed as modalities to slow or stop neuroma growth (neuroma ‘control’). Kondziolka et al (1998) described the efficacy of radiosurgery in a large series of patients but their methodology had significant shortcomings (O’Donoghue et al 1999). Forster et al (1996), after a study of 29 neuromas over a median of 6.6 years, concluded that stereotactic radiosurgery was an effective alternative treatment that did not replace microsurgery.
- 7.6.5 The reduced dose of radiosurgery to 10 – 20 gray has markedly reduced the occurrence of radiation-induced neuropathy. The addition of micro multileaf collimators to linear accelerators has facilitated the treatment of irregular volumes of tumour with a better three dimensional dose conformity than the gamma knife. Brainstem dose-volume histograms can be used to estimate the rate of cranial neuropathy from acoustic neuroma surgery (Meeks et al 2000).
- 7.6.6 All patients who undergo SR or FSR must submit to serial scanning for the rest of their lives or until neuroma growth is seen. The long-term follow-up of these patients is the responsibility of the team delivering the radiation treatment but the actual ‘face to face’ contact and imaging could be carried out by local specialists.
- 7.6.7 No controlled studies exist to show SR or FRS are better than no treatment. SR has been used extensively in the treatment of a variety of ‘benign’ intracranial lesions and, with the appropriate constraints and safeguards, can be a safe and effective therapy.
- 7.6.8 Concern exists about treating benign lesions, such as acoustic neuromas, with radiation, especially in the absence of tissue diagnosis. The long-term risks of such complications as brainstem ischaemia, and injury to cranial nerves (such as the facial nerve) are uncertain. Malignant change in a schwannoma following radiation treatment has been documented (Thomsen et al 2000). A further report satisfies Cahan’s criteria for radiation-induced malignancy (Shamisa et al 2001, Bance et al 2001).
- 7.6.9 Surgical removal of neuromas which grow despite radiation treatment is technically difficult and associated with poorer patient outcomes, especially in relation to facial nerve function (Battista et al 2000). However, the sample size (12 patients) was small, and the patients were not operated upon by the authors themselves.
- 7.6.10 Brada et al (1999) warn against equating activity with progress. They caution that the fact that over 80,000 patients have been treated worldwide with stereotactic radiosurgery could

be no more than the uncontrolled spread on an unproven technique. However, evidence from long-term outcomes studies to underpin this view was not provided.

8. OUTCOMES OF ACOUSTIC NEUROMA TREATMENT THE EVIDENCE BASE

- 8.1 The quality of evidence of articles relating to outcomes of acoustic neuroma management was evaluated. The widely accepted classification of the categories of evidence is shown in Table 1.
- 8.2 The search was confined to English language publications using the National Library of Medicine 'Medline' electronic retrieval system from 1977 – 2000. Only publications dealing with *outcomes from treatment* and with a *patient series of greater than 100 patients* were included.

TABLE 1: Classification of the Quality of Evidence

TYPE I	Evidence based on well-designed randomised controlled trials, meta-analyses or systematic reviews.
TYPE II	Evidence based on well-designed cohort studies or case control analytic studies.
TYPE III	Evidence based on well-designed non-experimental descriptive studies, such as comparative studies and correlation studies.
Type IV	Evidence based on expert committee reports, clinical experience of respected authorities, case reports, or on studies that have methodology problems such as sample size, length of follow-up, conflict in evidence.

TABLE 2: Quality of Published Evidence on the Outcome of Acoustic Neuroma Management

REFERENCE PAPER	TYPE OF STUDY	CASES	MAIN CONCLUSION	EVIDENCE TYPE
Hardy et al. J Neurosurg 1989; 71(6): 799-804	Surgery: translabyrinthine approach outcomes	100	Complete removal in 97% of cases, facial nerve anatomically intact in 82%	III
Lunsford et al. Otolaryngol Clin North Am 1992;25(2):471-91	Radiosurgery: outcomes	102	96% tumour control (1,7 years mean follow-up). It is an additional weapon and not replacement to surgery	IV
Cerullo et al. Surg Neurol 1993; 39(6): 485-93	Surgery: outcomes	102	Facial nerve function preserved in 86%, recurrence rate 10%	III

REFERENCE PAPER	TYPE OF STUDY	CASES	MAIN CONCLUSION	EVIDENCE TYPE
Robinette et al. Am J Otol 1997; 18(6): 378-45	Surgery: hearing preservation	104	Preoperative word recognition is significant predictor	III
Nutik SL. Surg Neurol 1994; 41(1): 28-33	Surgery: facial nerve outcomes	108	Size, experience and ease of dissection important factors	III
Nissan et al. Ear Nose Throat J 1997; 76(1): 37-40	Surgery: facial nerve outcomes	111	Size and approach not significant	III
Nissan et al. Laryngoscope 1997; 107(1): 118-21	Surgery: KTP-532 laser	111	KTR-532 laser is safe and has specific advantages	III
Fucci et al. Am J Otol 1999; 20(4): 495-9	No treatment – radiologic follow-up	119	Most neuromas with periodic MRI do not grow	III
Charabi et al. Otolaryngol Head Neck Surg 1995;113(1):5-14	No treatment – radiologic follow-up	123	Mean follow-up of 3.4 years, 18% of cases showed no growth; 8% smaller	III
Thomsen et al. Tokai J Exp Clin Med 1994; 19(3-6): 93-101	Surgery: algorithm	127	Management algorithm and 74% of neuromas grew with a mean of 3.4 mm/year and 75% lost hearing preservation candidature while waiting	III
Lalwani et al. Am J Otol 1995; 16(6): 758-64	Surgery: delayed facial nerve worsening	129	29% of patients experience delayed facial nerve worsening but with excellent prognosis	III
Lalwani et al. Otolaryngol Head Neck Surg 1994;111(5): 561-70	Surgery: facial nerve outcomes	129	90% of patients H-B scale I or II and the size of neuroma is important	III
Baguley et al. J Laryngol Otol 1992; 106(4): 329-31	Surgery: tinnitus	129	Post-operative tinnitus does not have a significant impact	III
Chung et al. Am J Otol 1997; 18(4): 436-43.	Surgery: quality of life	130	Surgery has not a considerable impact on patients' life	III
Rigby et al. Am J Otol 1997; 18(4): 427-35	Surgery: quality of life	130	Most symptoms are attributable to neuroma and not surgery	III
Van Leeuwen et al. Ann Otol Rhinol Laryngol 1996; 105(6): 423-30	Surgery: quality of life	134	Approach and size correlate with the outcome	III
Andersson et al. Am J Otol 1997;18(4):421-6	Surgery: quality of life	141	Few patients experience social problems post-operatively	III
Andersson G. J Psychosom Res 1999; 46(3):257-60	Surgery: quality of life	141	Anxiety, age and facial nerve function were associated with symptoms	III
Van Roijen et al. Acta Neurochir (Wien) 1997; 139 (1):942-8.	Surgery versus radiosurgery	145	Radiosurgery more cost-effective	IV
Slattery et al. Am J Otol 1997;18(5):596-601	Surgery: middle fossa approach	151	Minimal complications-good to excellent results	III
Dutton, Ramsden et al. J Laryngol Otol 1991;105(3): 165-173.	Surgery: outcomes	151	Mortality 3%, good or normal facial nerve 79%. Size and experience significant.	III
Henrich et al. Ear Nose Throat J 1995;74(7):462-6	Surgery: tinnitus	160	75% of patients report post-operative tinnitus	III
Cohen et al. Am J Otol 1993;14(5):423-33	Surgery: hearing preservation	161	Complications were somewhat increased by attempted such surgery	III
Glasscock et al. J Neurosurg 1993; 78(6): 864-70	Surgery: hearing preservation	161	Hearing preservation achievable in 35% of cases	III

REFERENCE PAPER	TYPE OF STUDY	CASES	MAIN CONCLUSION	EVIDENCE TYPE
Kondziolka et al. N Engl J Med 1998; 339(20): 1426-33	Radiosurgery: long term outcomes	162	Radiosurgery provides long term control with minimal complications	III
Arriaga et al. Am J Otol 1994;15(5):620-4	Surgery: comparison of approaches	164	Translab and middle fossa similar facial nerve outcomes for neuromas < 1.5 cm	III
Briggs et al. Neurosurgery 1994;34(5):785-90	Surgery: translab removal of large neuromas	167	Acceptable facial nerve function in 75%, complications 4-10%	III
Gormley et al. Neurosurgery 1997;41(1): 50-8	Surgery: outcomes	179	Surgery is preferred over radiosurgery	III
Gormley et al. Neurosurgery 1997; 41(1): 50-60.	Surgery: outcomes	179	Surgery is preferred over radiosurgery in healthy patient	III
Falcioni et al. Am J Otol 1999; 20(5): 660-6	Surgery: complications	200	With proper technique 0% of CSF leak	III
Driscoll et al. Am J Otol 19(4): 491-5	Surgery: complications	210	31% of patients experience post-operative disequilibrium	III
El-Kashlan et al. Am J Otol 1998; 19(1): 104-11	Surgery: vestibular symptoms	220	Significant correlations between pre-op symptoms and vestibular test results and post-op dizziness disability	III
Lynn et al. Am J Otol 1999; 20(4): 484-94	Surgery: complications	237	65% of patients have disequilibrium but with low impact on their life	III
Noren et al. Acta Neurochir Suppl (Wien) 1993;58:104-7	Radiosurgery: outcomes	254	Tumour control in 88%, hearing preservation 77%, trigeminal neuropathy 19%	III
Irving et al. Laryngoscope 1995; 105(8pt1): 809-13	Surgery: complications	257	Nervus intermedius abnormalities are common	III
Irving et al. Am J Otol 1995; 16(3): 331-7	Surgery: quality of life	257	Excellent post-operative quality of life	III
Mass et al. Arch Otolaryngol Head Neck Surg 1999; 125:801-4	Surgery: complications of translabyrinthine approach	258	Safe and effective approach	III
Grey et al. Clin Otolaryngol 1996; 21(5): 409-413	Surgery: translabyrinthine versus retrosigmoid	276	Retrosigmoid give better facial nerve outcomes	III
Paring et al. Arch Otolaryngol Head Neck Surg 1992; 118(10): 1061-4	Surgery: quality of life	293	Deafness, balance and facial nerve cause the most severe problems post-operatively	III
Symon et al. Br J Neurosurg 1989; 3(3): 343-7	Surgery: outcomes	392	Good or excellent result in 94%, mortality rate 1.4%,	III
Koos et al. J Neurosurg 1998; 88(3): 506-12	Surgery: hearing preservation	442	Hearing preservation from 57% to 100% of patients according to size and location	III
Niranjan et al. Neurosurg Clin N Am 1999;10(2):305-15	Radiosurgery: hearing improvement	487	21 patients out of 487 experienced hearing improvement	III
Niranjan et al. Neurosurg Clin N Am 199; 10(2): 305-15	Radiosurgery: hearing improvement	487	In 4.3% of cases hearing improved	III

REFERENCE PAPER	TYPE OF STUDY	CASES	MAIN CONCLUSION	EVIDENCE TYPE
Arriaga et al. Otolaryngol Head Neck Surg 1993; 108(3): 220-4	Surgery: facial nerve outcomes	515	Acceptable facial nerve function over 90% of cases	III
Wiegand et al. Laryngoscope 1989; 99(2): 179-87	Surgery: quality of life	541	64% of respondents had some facial weakness and there is a great need for information or patient orientated care	III
Selesnick et al. Am J Otol 1998; 19(6): 846-9	No treatment – radiologic follow-up (meta-analysis)	571	Mean follow-up of 3 years, 46% of cases showed no growth	III
Guerin et al. Ann Acad Med Singapore 1999; 28(3): 402-8	Surgery: complications	611	Surgery is the best treatment for the majority of cases	III
Sampath et al. J Neurosurg 1997; 87(1): 60-6	Surgery: facial nerve outcomes	611	90% of cases House-Brackmann 1 or 2	III
Samii et al. Adv Tech Stand Neurosurg 1995; 22:343-73	Surgery: hearing preservation	653	Hearing preservation rate was 51% in small neuromas and 22% in large neuromas	III
Samii et al. Neurosurgery 1997; 40(1): 11-21.	Surgery: complications	1000	Sub occipital approach and careful selection should reduce mortality rate to <1%	III
Samii et al. Neurosurgery 1997; 40(4): 684-94	Surgery: facial nerve outcomes	1000	Anatomical preservation of facial nerve in 93%	III
Samii et al. Neurosurgery 1997; 40(2): 248-60	Surgery: hearing preservation rate	1000	Hearing preservation is achievable (40% of cases)	III

8.3 The Evidence Base – Conclusions

- 8.3.1 A number of neuromas involute or do not exhibit further growth after initial diagnosis. Thus, the available evidence supports the strategy of watchful waiting with interval scanning in selected cases. No predictors exist that can consistently identify those neuromas that will subsequently increase in size.
- 8.3.2 Evidence demonstrates that surgery can achieve total neuroma removal in up to 97% of patients with mortality, in centres reporting results, of approximately 1%.
- 8.3.3 Some degree of facial paralysis complicates neuroma removal in a significant number of patients and represents a major source of disability.
- 8.3.4 Radiotherapeutic approaches can achieve a ‘control’ rate in a significant number of patients, at least in the short term. Long-term risks, including malignant change and injury to adjacent neural structures, remain a concern. Life long surveillance is needed following treatment.
- 8.3.5 Almost all the above studies assessed each treatment modality separately, were generally retrospective, and thus represent a less than compelling level of evidence (Level III or IV in Table 1).
- 8.3.6 No study exists that systematically compares the different modalities of management (surgery, radiosurgery, interval scanning).

9. QUALITY OF LIFE FOLLOWING ACOUSTIC NEUROMA SURGERY

- 9.1 Quality of life studies comparing post-operative quality of life with pre-operative status need to be interpreted with caution. When most patients present with an acoustic neuroma, they enjoy excellent health. They thus opt for surgery with the aim of preserving life and preventing major neurological complications, possibly many years later.
- 9.2 Another important shortcoming in assessing quality of life and comparing the different modalities of treatment is the lack of standardised and validated instruments that can reliably measure and compare the quality of life in patients with acoustic neuroma across the various methods of management.
- 9.3 Using a validated health-status instrument (The Glasgow Benefit Inventory), it was found that 54% of UK patients experienced a poorer quality of life after surgery as compared to their pre-operative status (Nikolopoulos et al 1998). Fifty per cent engaged in fewer social activities after surgery. Surgery had a greater impact on the life quality of younger patients. A statistically significant correlation between quality of life and neuroma size was not found. These results are similar to those reported by a Danish Study (Jorgensen and Petersen 1994) but at variance with a study by Irving et al (1995) and Chung et al (1997) who found surgery had minimal impact on life quality.
- 9.4 A further UK study (Bateman et al 2000), using open-ended questionnaire techniques, categorised patients' symptoms according to the World Health Organisation's classifications of impairment, disability and handicap. The study revealed a wide variety of post-operative symptoms, especially visual disturbance (49% of patients) and psychosocial deficits (34% of patients)
- 9.5 A Dutch study found 25% of surgical patients were declared unfit for work following surgery compared with an American study showing that only 1.6% became unemployed after surgery (Van Leeuwen et al 1995 & 1996, Chung et al 1997). However it is clear that the recovery time following stereotactic radiosurgery is shorter than that after microsurgery. Those patients in the surgical group with paid jobs were absent from work for 3 months on average. Patients with jobs undergoing stereotactic radiosurgery were absent for one working day (Van Roijen et al 1997).
- 9.6 One UK study of a consecutive series of surgically treated patients found that almost 80% of patients continued without change with their usual occupations (Nikolopoulos et al 1998).
- 9.7 Patients and surgeons do not always agree about outcomes, especially concerning facial nerve function following surgery (Wiegand et al 1989). Nevertheless, an objective independent assessment of facial nerve function, for example using the House Brackmann scale, can be a useful index of this specific neurological limitation following different methods of management.
- 9.8 Cross et al (2000), using 4 validated questionnaires, found that the distress experienced by patients with facial paralysis following acoustic neuroma surgery does not correlate with the grade of the paralysis as attributed by surgeons. They found that even patients with minimal disturbance of facial nerve function may experience great personal distress (Cross et al 2000).

10. COST – EFFECTIVENESS

- 10.1 No formal studies exist that compare the cost-effectiveness of the various treatment options in acoustic neuroma patients.
- 10.2 One Dutch study compared the costs of microsurgery with those of stereotactic radiosurgery (Van Roijen et al 1997). This demonstrated that the direct costs of surgery exceeded those of radiosurgery by 20%. However, the outcomes of these two treatment modalities are not equivalent. Surgery results in total neuroma removal whereas radiosurgery does not and requires the patient to undergo life-long serial scanning.

11. INDICATORS OF GOOD PRACTICE

- 11.1 The care of acoustic neuroma patients requires multidisciplinary teamwork with access to the full range of specialist support services these patients may need.

Centres offering this care should have:

- 11.2 An otolaryngologist and a neurosurgeon with a specialist interest and training in neuro-otology and skull base surgery as well as access to specialist facilities in stereotactic neurosurgery.
- 11.3 There should be links and communication between those neurosurgeons with specialist expertise in stereotactic radiation therapy (at a supraregional or national level) able to contribute both to the clinical management in broad terms as well as recommend and supervise intervention by stereotactic radiation therapy.
- 11.4 Audiological facilities to undertake diagnostic auditory and vestibular investigations as well as post-operative auditory and vestibular rehabilitation. Post-operative dizziness and tinnitus can be troublesome and patients may need supportive therapy. The provision of CROS hearing aids should be considered in appropriate patients. Referral to an audiological physician may be desirable in the rehabilitation of certain patients.
- 11.5 Neurodiagnostic imaging and neuroradiological support, including facilities for emergency imaging.
- 11.6 Neuroanaesthetic provision consistent with standards described in ‘Guidance on the Provision of NeuroAnaesthesia’ (pages 60-65) in Guidelines for the provision of Anaesthetic Services published by the Royal College of Anaesthetists - July 1999.
- 11.7 Neuromonitoring facilities for monitoring the facial nerve and, where indicated, hearing function during surgery.
- 11.8 Access to intensive care facilities.
- 11.9 Patient numbers: the centre should have sufficient level of clinical activity to gain familiarity with the diverse needs of acoustic neuroma patients as well as to develop and maintain surgical skills and provide surgical training.
- 11.10 Audit: regular multidisciplinary audit of treatment outcomes.

- 11.11 Plastic and Reconstructive Surgery: access to these facilities, especially for facial reanimation surgery. Access to oculoplastic surgical expertise may be helpful in the management of eye-lid dysfunction.
- 11.12 Referral to voluntary support agencies when requested.

12. THE PATIENTS' PERSPECTIVE

- 12.1 A number of patient organisations, such as the British Acoustic Neuroma Association, offer a network of support for patients. Patients should be made aware of the existence of these organisations and should be helped to contact them if they so chose.
- 12.2 The RNID is the largest charity representing the 8.7 million deaf and hard of hearing people in the UK. It supports initiatives which provide evidence-based information to empower people to make informed choices. The RNID therefore welcomes the production of these guidelines as a means of assisting professionals and patients in decisions relating to the diagnosis and management of acoustic tumours to improve the quality and consistency of patient care.

13. AUDIT AND CLINICAL GOVERNANCE

- 13.1 A national audit of acoustic neuroma treatment results has not been undertaken.
- 13.2 A national audit would enable units to compare their performance across patient groups and would help the implementation of clinical governance in acoustic neuroma practice. However, validated clinical outcomes need to be developed to ensure meaningful comparisons are made across treatment modalities. It would also facilitate the accrual of sufficient numbers to compare treatments in a prospective manner with sufficient statistical power.
- 13.3 With agreement between teams on a minimum data set and with appropriate administrative support, such studies could be readily undertaken.
- 13.4 Centres offering SR and FSR should be limited in number and identified nationally. Appropriate criteria and a process of designation need to be developed. They should offer life-long surveillance and make available their results for treating acoustic neuroma patients, especially their long-term outcomes.

14. EQUITY OF ACCESS

- 14.1 All patients with acoustic neuromas should have access to a uniformly high standard of care. Evidence suggests that patients achieve the best outcomes in centres with a special interest in this condition. Therefore, patients should be referred to a specialist unit known to have the expertise in acoustic neuroma management (which may not be the nearest hospital).

15. HEALTH PROMOTION

- 15.1 Greater public awareness of the importance of unilateral or asymmetrical auditory symptoms would do much to facilitate earlier diagnosis of acoustic neuromas. '*Its time to test your hearing*' by the Royal National Institute for Deaf People (RNID) is an example of a programme which aims to educate the public about hearing disorders. The RNID has also produced a fact sheet on acoustic neuroma.
- 15.2 General Practitioners (GP's). The first point of contact with the health care system for most patients remains the family practitioner. Continuing Education programmes should remind GP's of the importance of referring patients who present with unilateral or asymmetrical auditory symptoms to their local ENT Department. The introduction of hearing testing into GP Health Screening would be particularly helpful.
- 15.3 When patients are referred directly to audiology departments, protocols for onward referral of those patients with findings suggestive of acoustic neuromas should be implemented.

16. UPDATING

These guidelines will be updated in March 2003.

17. REFERENCE LIST

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**Clinical Effectiveness Guidelines
Acoustic Neuroma
(Vestibular Schwannoma)**

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British Association of Otorhinolaryngologists – Head and Neck Surgeons
British Society of Audiology
British Society of Neuroradiologists
British Skull Base Society
Royal National Institute for the Deaf People
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Disclaimer

The views represented in these guidelines are not necessarily the views of all members of the working party or their Associations. They are intended as a general guide to patient management and should not replace the clinical judgement professionals should exercise in managing individual patients.

Main Recommendations

1. Acoustic neuromas should be suspected in patients who present with unilateral or asymmetrical auditory symptoms (hearing loss or tinnitus). Magnetic resonance imaging represents the method of choice for identifying the minority of these patients who have an underlying acoustic neuroma.

2. Patients who are found to have an acoustic neuroma should be presented with the relevant management options available to them when they are initially diagnosed. Information about voluntary support agencies should be made available to patients.

3. The management options currently available include interval scanning, surgical removal or radiation treatment. The effect of each of these modalities is different and the selected strategy needs to be appropriate to the individual circumstances of the patient.

4. Management of these patients is best undertaken by dedicated multidisciplinary teams working in specialist centres.

5. There is need for prospective longitudinal outcomes research to compare the relative merits of the various management modalities, especially in those patients with small neuromas or where no tumour growth has been demonstrated.

6. A national audit of acoustic neuroma is needed to analyse outcomes across centres to ensure a uniformly acceptable standard of care is available to patients nationally. Validated measures that assess all aspects of outcomes (included quality of life issues) need to be developed and implemented in clinical practice.

1. Objectives

The purpose of these guidelines is as follows:

- 1.4 To assist professionals and patients about decisions relating to the diagnosis and management of acoustic neuromas based on the best available evidence. These guidelines may also be of value to health-care commissioners in their purchasing decisions.
- 1.5 To facilitate the early detection of acoustic neuromas.
- 1.6 To improve the quality and consistency of patient care and outcome across Great Britain and Ireland.

Note: The Guidelines do not cover the care of patients with Neurofibromatosis II or those patients with non-acoustic neuromas.

2. Introduction

- 2.3 Acoustic neuromas represent about 6% of all intracranial tumours. They are benign, slow-growing tumours, which arise from cells in the sheaths that surround the hearing and balance nerves. The neuromas usually manifest themselves as one-sided hearing impairment, which may go ignored by the patient or be dismissed by the doctor. Continued growth of these neuromas ultimately results in compression on the brainstem and a rise in pressure within the brain (raised intracranial pressure). Early diagnosis offers patients a range of management options and may significantly reduce morbidity.
- 2.4 Pathologically, acoustic neuromas are vestibular schwannomas. Because the term 'acoustic neuroma' is in such common usage, this term has been maintained throughout the text.

4. Most Frequent Presenting Symptoms

- 3.1 About 90% of patients present with gradual progressive hearing loss in one ear. However, about 5% will present with a sudden hearing loss. Many (about 70%) experience tinnitus (noise) in one ear. However, about 3 % of patients with acoustic neuromas will have normal hearing at presentation. Patients, especially when asked, will often admit to experiencing balance disturbance, which may be quite mild.
- 3.2 Less common presentations include facial numbness or pain from involvement of the trigeminal nerve, inco-ordination from cerebellar compression or earache. Facial weakness is very uncommon despite the fact that the neuroma is pressing on and causing damage to the facial nerve. Large or medially-placed neuromas may have symptoms of raised intracranial pressure such as headaches, visual disturbance or a diminution in the level of responsiveness.
- 3.4 The advent of Magnetic Resonance Imaging (MRI) has resulted in the diagnosis of symptomless acoustic neuromas in patients who were scanned for other unrelated conditions (Selesnick et al. 1993).

4. Epidemiology of Acoustic Neuroma

- 4.9 The incidence of acoustic neuromas - the number of newly diagnosed cases per year – is around 13 cases/million/year (Moffat et al 1989, Davis 1995). Tos et al (1999) have shown that incidence figures in Denmark have increased over the last 20 years from 7.8 to 12.4 cases/million/year. This is probably a reflection of better diagnostic methods rather than a true increase in neuroma incidence.
- 4.10 These incidence figures are based on patients who have received a diagnosis whereas estimates of prevalence — the number of cases in the population — have so far been estimated only on the basis of large, unselected autopsy or radiology studies.
- 4.11 An autopsy study by Leonard and Talbot (1970), suggested a prevalence of 0.8 % or 8,000 per 1,000,000. If one accepts the clinical incidence of acoustic neuromas as being between 13 per 1,000,000, it is reasonable to assume that many neuromas escape diagnosis.
- 4.12 A recent radiological study found 7 unsuspected acoustic neuromas per 10,000 brain MRI studies - equivalent to 700 cases per 1,000,000 population (Anderson et al 2000).
- 4.13 The number of patients with asymmetrical hearing symptoms, who attend ENT clinics and are eventually diagnosed with acoustic neuroma, ranges from 3-7.5% (Ferguson et al 1996; Hollingworth et al 1998).
- 4.14 Within a District General Hospital ENT clinic it has been shown that 19.7% of new patients attending the ENT outpatient department are potential candidates for screening for acoustic neuromas (Harcourt et al 1999).
- 4.15 The National Study of Hearing showed that 2.9% of the population has an asymmetry greater or equal to 15 dB across 0.5 – 4 kHz. For the high frequencies 4, 6, and 8 kHz, this prevalence increases to 10.4% (Davis et al 2000). Where the better ear has hearing thresholds better than 25 dB, the prevalence values are 5.2% and 10.9% respectively.
- 4.16 These data highlight the need for effective screening of patients with asymmetrical hearing symptoms, as only a minority will prove to have an acoustic neuroma.

5. Who should be investigated?

- 5.3 The diagnosis of acoustic neuroma should be considered in any patient complaining of unilateral or asymmetrical auditory symptoms (either hearing loss or tinnitus), whether of progressive or sudden onset in whom there is no other obvious cause. The diagnosis should also be considered in patients with impaired facial sensation and in certain patients with imbalance, which cannot otherwise be explained.
- 5.4 Age alone should not dictate whether or not a patient with appropriate symptoms is investigated. Knowledge of the diagnosis, even if no treatment is offered, may help in patient management.

6. What investigations are appropriate?

6.1 Audiological

- 6.1.1 **Pure Tone Audiometry**
All patients with unilateral or asymmetrical hearing symptoms should have a pure tone audiogram, appropriately masked as necessary. If previous audiometry is available, any change in threshold should be noted. Most patients have a high frequency sensorineural loss at presentation but any pattern of hearing loss (or normal hearing) may be encountered.
- 6.1.2 **Speech Discrimination Testing**
In the past, much emphasis was placed on speech audiometry for 'site of lesion' testing. However, it has largely been supplanted in this role. The test is still of great value to assess the usefulness of hearing in the neuroma ear, especially when hearing conservation surgery is being considered.
- 6.1.3 **Electric Response Audiometry**
Auditory brainstem responses (ABR) have been described as having high sensitivity and specificity (over 90%) in the diagnosis of acoustic neuromas (Hood 1998). However, careful determination of the false negative rate of ABR, indicates that especially for small neuromas it is unacceptably high and approaches 17% (Wilson 1997). Its usefulness is further limited by the fact that in cases of severe to profound hearing loss, no ABR response can be elicited due to the extent of the hearing loss. An absent ABR in the presence of milder losses, however, is suggestive of retro-cochlear pathology. There has been recent interest in the use of otoacoustic emissions to help assess the status of the cochlea in patients with acoustic neuromas.
- 6.1.4 **ABR represents the non-imaging test of choice in the small number of patients in whom MRI may be contraindicated or not tolerated.** The test is non-invasive, is well tolerated by patients, but requires sophisticated equipment and a high degree of expertise, especially in the interpretation of test results. Open MRI may also be an option for such patients (but the field strength of open magnets is significantly reduced and may be inadequate for screening purposes).
- 6.3 **Diagnostic Imaging**
- 6.3.1 **Computed Tomography (CT)**
CT scanning is performed using thin sections (2–3 mm) after intravenous injection of an iodine-containing contrast medium and filmed at soft tissue and bone window settings. It has the advantage of being widely available, cheaper than magnetic resonance imaging (MRI) and it shows bone erosion of the internal auditory canal (IAC) to best advantage. Some patients who cannot be examined by MRI (claustrophobia, cardiac pacemaker etc.) may be scanned by CT.
- 6.3.2 **CT provides excellent visualisation of moderate or large (>15 mm) neuromas in the cerebello-pontine angle cistern with a sensitivity of detection approaching 100%.** However, the sensitivity falls if the neuroma is smaller than 10 mm and CT is unable to detect those neuromas entirely confined to the IAC (intra-canalicular neuromas).
- 6.3.3 **In the majority of cases it is highly desirable to detect acoustic neuromas when they are small.** This lack of sensitivity and the modest dose of ionising radiation imparted to the patient make CT an inappropriate method of screening large numbers of patients. However, depending on local availability and cost of MRI, CT may retain a role in scanning elderly patients where the aim is limited to detecting larger space-occupying lesions.

6.2.4 **Magnetic Resonance Imaging (MRI)**

MRI is the most accurate diagnostic test for identifying acoustic neuromas (Curtin 1997). It also has the advantages of multiplanar imaging, of enabling an assessment of the labyrinth, and it does not involve ionising radiation. It has largely supplanted CT in the investigation of patients with unilateral or asymmetrical audiovestibular symptoms. The two major approaches for screening patients in order to detect acoustic neuromas with MRI use T2-weighted images or contrast enhanced T1-weighted images.

6.2.5 One strategy is to acquire T2-weighted images only. The spatial resolution of the images must be sufficient to reliably demonstrate the cisternal and intracanalicular portions of the VII and VIII nerves, such that for small neuromas it is possible to identify which branch of the vestibulocochlear nerve (or rarely, the facial nerve) is involved. T2-weighted images also enable identification of whether there is residual CSF between the neuroma and the fundus of the IAC, which may influence surgical planning. Vascular compressive lesions, peritumoral oedema and associated arachnoid cysts are also revealed. Cochlear anomalies and large vestibular aqueduct syndrome, which may account for as many as 10% of abnormalities associated with sensorineural hearing loss, are easily identified (Daniels et al 2000). Although some intralabyrinthine neuromas can be visualised, very small neuromas may evade detection.

6.2.7 If T2-weighted images are to be performed, every care should be taken to optimise scan parameters. Employing a standard 2D FSE sequence with a slice width of 3 mm slice runs the dual risk of incurring a high false negative rate and having a high rate of equivocal scans requiring additional T1-weighted enhanced images (Zealley et al 2000). Scanner capabilities vary so much that it is impractical to specify parameters. However, close attention needs to be paid to slice thickness (maximum 2 mm), slice interleaving, matrix size (e.g. 512 x 512), number of acquisitions (e.g. 4-6), and field of view (maximum 20 x 20 cm) along with the possibility of using dual phased array receiver coils over both temporal bones. 2D images should be acquired in axial and coronal planes.

6.2.7 Scans should be interpreted by appropriately experienced neuroradiologists or ENT radiologists, and patients recalled for contrast enhanced imaging in cases of radiological uncertainty.

6.2.8 However, if available, a single acquisition using a T2-weighted 3D FSE or T2*-weighted CISS technique offers considerable advantages in spatial resolution (slice thickness <1 mm) with potential for image reconstruction and slice overlap to show the nerves and labyrinthine structures in greater detail. High resolution 2D or 3D T2-weighted techniques, in conjunction with appropriate clinical pre-screening and referral, can provide a very sensitive method of evaluating unilateral sensorineural hearing loss compared to contrast-enhanced T1-weighted images (Daniels et al 1998 & 2000). It is also advisable to perform a T2-weighted FSE (or similar) sequence through the brain especially if the brainstem has not been well visualised by the focussed high resolution images.

6.2.8 As only a limited number of sequences are employed, examination times are short. Medical staff need not supervise the scans and if suitable patients are "batched", large numbers can be scanned in a given MRI session. However, this approach may not be suitable for older generation or lower field (< 1T) scanners where spatial resolution is insufficient to clearly define the individual nerves. It may prove necessary in a small percentage of patients to obtain additional contrast-enhanced T1-weighted images patients with equivocal findings on T2-weighted images or where patient movement leads to an inability to resolve the individual

components of the nerve complex. For scans that are medically unsupervised, this requires patients to be recalled. It should be recognised that radiologists reporting these scans will have varying degrees of training and familiarity with imaging this region, and some may have a preference for and greater confidence with reporting contrast-enhanced T1-weighted images.

- 6.2.9 The other approach is to only acquire T1-weighted images. Injection of a gadolinium-based contrast medium increases the sensitivity of detecting small neuromas and intralabyrinthine lesions, non-enhancing schwannomas being extremely rare. It is considered by most to be the gold standard for detecting acoustic neuromas. Inflammatory disorders of the VIII nerves and labyrinths may also be revealed. The anatomical detail of the nerves and fluid containing structures however is much poorer than on T2-weighted images.
- 6.2.10 T1-weighted images are acquired in the axial plane, and often the coronal plane, using 2-3 mm sections. Preliminary unenhanced T1-weighted images may help detect rare lesions such as lipoma or labyrinthine haemorrhage but many centres in the UK routinely acquire T1-weighted images only after intravenous injection of contrast medium. The gadolinium-containing contrast media are well tolerated with an extremely low incidence of side effects.
- 6.2.11 Injecting a contrast agent increases overall examination time and requires medical staff to inject or be in close proximity if it is to be administered by radiographic staff. This may preclude scheduling of scans outside normal working hours. The use of contrast medium increases the cost of the examination although it is not necessary to inject as much as 0.1 mmol/kg. Five-ten ml of contrast agent is sufficient, limiting the additional expense to £20-30.
- 6.3.12 In centres with a substantial otolaryngology service, MRI requests for patients with suspected acoustic neuromas might comprise 10% or more of all MRI examinations. Whilst T1-weighted and T2-weighted images can provide complimentary information, the routine use of both techniques may be difficult to justify for screening purposes in the face of limited resources. Selecting the appropriate screening protocol will depend upon local circumstances (e.g. type of scanner, MRI session availability, intravenous (I.V). injection protocols, radiologist's skills and preferences) and upon balancing the small risk of missing a very small intracanalicular or intralabyrinthine neuroma against cost and increasingly long waiting times for investigation and diagnosis. There is an increasing consensus that T2-weighted imaging, if of a sufficiently high quality, can be employed as a highly accurate and cost effective screening test where the overall demand for MRI services is very high (Daniels et al 2000).

7. Management Options

- 7.1 There are three management options for acoustic neuroma patients:
 - a. Interval scanning,
 - b. Surgical removal of the neuroma or
 - c. Stereotactic radiosurgery / stereotactic fractionated radiotherapy
- 7.2 The major determinants of which treatment is adopted are: neuroma size, age, health-status, the desire to attempt hearing preservation, the state of hearing in the opposite ear, and the preference of the patient after due consideration of the advantages and risks of each option.

7.3 Patients should be presented with the full range of management options available to them when they present.

7.4.8 **No intervention with interval scanning**

7.4.9 This strategy may be considered, at least in the short term, for patients with a small neuroma and good hearing. Older patients and individuals in poor health may be managed in this way, certainly initially, although it is by no means certain that neuromas will grow less aggressively in old people.

7.4.10 For intracanalicular neuromas, an observation period between scans of approximately 1 year may be appropriate as there is evidence that some of these neuromas cease to grow, at least in the short term. (O'Reilly et al 2000).

7.4.11 Spontaneous involution of acoustic neuromas has been reported in up to 13% of patients (Luetje 2000).

7.4.12 In one study, two-thirds of neuromas did not grow over a mean follow-up period of 35 months and neuroma involution was observed in 12% of cases. Growth in the first year following diagnosis was predictive of later neuroma enlargement. The authors recommended that in the absence of documented growth, watchful waiting represent the best management option. (Tschudi et al 2000).

7.4.13 Evidence suggests that, using the middle fossa approach, a hearing conservation rate of 69% can be achieved in small neuromas with good pre-operative hearing (Weber et al 1996). Allowing such neuromas to grow, especially in young patients, could compromise the ability to preserve hearing.

7.4.14 All patients being managed conservatively should be reviewed by annual imaging, to look for neuroma growth. Neuromas demonstrating growth (as demonstrated on serial scans by an increase in cross-sectional diameter or by an increase in neuroma volume) should then be considered for either surgery or radiotherapy. However, there is no agreed, validated measure of neuroma size across centres.

7.5 **Surgical Removal**

7.5.1 Surgical removal is the treatment offered to the majority of patients with acoustic neuromas. Two surgical approaches predominate in the UK: the retrosigmoid (RS) and translabyrinthine (TL). Other less frequently employed operations are the Middle Fossa (MF), the Extended Middle Fossa (EMF) and Transotic approaches.

7.5.2 The RS, MF and EMF approaches provide the chance of preserving residual hearing in a subset of patients with good pre-operative hearing and suitably located neuromas – the chances of succeeding in this particular subgroup of patients being no greater than 50%. Hearing conservation should also be considered in patients who have poor hearing in the contralateral ear. There are no agreed audiological criteria for hearing conservation. The RS or MF approach is therefore employed for patients with useful residual hearing and a reasonable expectation of hearing preservation. The TL is generally employed for patients with poor hearing and /or larger neuromas where hearing preservation is not achievable.

- 7.5.8 Complete neuroma removal is achievable in over 95% of cases (Hardy et al 1989).
- 7.5.9 Operative mortality in experienced centres is less than 1% with the risk being slightly greater with larger neuromas (Ramsden 1995). A possible risk of epilepsy attends middle fossa approaches.
- 7.5.10 Permanent facial paralysis, either partial or complete, remains the greatest single source of disability following neuroma removal; those patients with large neuromas are at greatest risk of this complication (Cerullo et al 1993; Lalwani et al 1994; Nutik et al 1994). Other factors, such as the degree of adherence and intermingling between tumour and nerve, make precise pre-operative prediction difficult in individual patients. The most commonly used clinical grading system for facial paralysis, the House Brackmann Scale (House et al 1985), should be used in reporting of results.
- 7.5.11 A learning curve has been described in acoustic neuroma surgery with a plateau being reached between 20 and 60 cases (Welling et al 1999, Buchman et al 1996). A surgeon commencing acoustic neuroma surgery should be appropriately trained, preferably having carried out an appropriate number of operations under supervision prior to establishing an independent practice. Surgery is often undertaken as a collaborative exercise between the neuro-otologist and the neurosurgeon, especially for large neuromas.
- 7.5.12 The use of facial nerve monitoring has improved the outcome of patients undergoing acoustic neuroma surgery and its use is considered mandatory for any acoustic neuroma operation (Kartush 1998). Monitoring cochlear function may also be useful in hearing conservation surgery.

7.6 Stereotactic radiosurgery (SR) and fractionated stereotactic radiotherapy(FSR)

- 7.6.11 Stereotactic radiosurgery (SR) was defined by its pioneers (Leskell and Larsson) as the application of single fraction ionising radiation to a stereotactically defined volume of tissue, irrelevant of its histological composition.
- 7.6.12 More recently, techniques have been refined to enhance treatment of the neuroma and reduce the risk of radiation damage to surrounding structures. This can be achieved by reducing the dosage of radiation, using a stereotactic technique to treat irregular lesions in their three dimensions, and giving multiple small doses - fractionated stereotactic radiotherapy (FSR). Image fusion to optimise shaping and planning should be a standard of care if radiotherapeutic approaches are used. Small and medium sized neuromas up to 3.0cm in diameter are considered as being potential candidates for SR or FSR treatment (Forster et al 1996).
- 7.6.13 The source of radiation in SR and FSR is either gamma ray photons from multiple high activity Cobalt-60 sources or a linear accelerator (LINAC) which uses X-ray photons derived from high-energy electrons. Although both sources can be used for SR and FSR, in practice the Cobalt source is almost exclusively used to deliver SR.
- 7.6.14 SR and FSR do not remove neuromas but are generally proposed as modalities to slow or stop neuroma growth (neuroma 'control'). Kondziolka et al (1998) described the efficacy of radiosurgery in a large series of patients but their methodology had significant shortcomings

(O'Donoghue et al 1999). Forster et al (1996), after a study of 29 neuromas over a median of 6.6 years, concluded that stereotactic radiosurgery was an effective alternative treatment that did not replace microsurgery.

- 7.6.15 The reduced dose of radiosurgery to 10 –20 gray has markedly reduced the occurrence of radiation-induced neuropathy. The addition of micro multileaf collimators to linear accelerators has facilitated the treatment of irregular volumes of tumour with a better three dimensional dose conformity than the gamma knife. Brainstem dose-volume histograms can be used to estimate the rate of cranial neuropathy from acoustic neuroma surgery (Meeks et al 2000).
- 7.6.16 All patients who undergo SR or FSR must submit to serial scanning for the rest of their lives or until neuroma growth is seen. The long-term follow-up of these patients is the responsibility of the team delivering the radiation treatment but the actual 'face to face' contact and imaging could be carried out by local specialists.
- 7.6.17 No controlled studies exist to show SR or FRS are better than no treatment. SR has been used extensively in the treatment of a variety of 'benign' intracranial lesions and, with the appropriate constraints and safeguards, can be a safe and effective therapy.
- 7.6.18 Concern exists about treating benign lesions, such as acoustic neuromas, with radiation, especially in the absence of tissue diagnosis. The long-term risks of such complications as brainstem ischaemia, and injury to cranial nerves (such as the facial nerve) are uncertain. Malignant change in a schwannoma following radiation treatment has been documented (Thomsen et al 2000). A further report satisfies Cahan's criteria for radiation-induced malignancy (Shamisa et al 2001, Bance et al 2001).
- 7.6.19 Surgical removal of neuromas which grow despite radiation treatment is technically difficult and associated with poorer patient outcomes, especially in relation to facial nerve function (Battista et al 2000). However, the sample size (12 patients) was small, and the patients were not operated upon by the authors themselves.
- 7.6.20 Brada et al (1999) warn against equating activity with progress. They caution that the fact that over 80,000 patients have been treated worldwide with stereotactic radiosurgery could be no more than the uncontrolled spread on an unproven technique. However, evidence from long-term outcomes studies to underpin this view was not provided.

9. Outcomes of Acoustic Neuroma Treatment - The Evidence Base.

- 8.1 The quality of evidence of articles relating to outcomes of acoustic neuroma management was evaluated. The widely accepted classification of the categories of evidence is shown in Table 1.
- 8.3 The search was confined to English language publications using the National Library of Medicine 'Medline' electronic retrieval system from 1977 - 2000. Only publications dealing with *outcomes from treatment* and with a *patient series of greater than 100 patients* were included.

Table 1: Classification of the Quality of Evidence

TYPE I	Evidence based on well-designed randomised controlled trials, meta-analyses or systematic reviews
TYPE II	Evidence based on well-designed cohort studies or case control analytic studies
TYPE III	Evidence based on well-designed non-experimental descriptive studies, such as comparative studies and correlation studies
Type IV	Evidence based on expert committee reports, clinical experience of respected authorities, case reports, or on studies that have methodology problems such as sample size, length of follow-up, conflict in evidence

Table 2: Quality of Published Evidence on the Outcome of Acoustic Neuroma Management

REFERENCE PAPER	TYPE OF STUDY	CASES	MAIN CONCLUSION	EVIDENCE TYPE
Hardy et al. J Neurosurg 1989; 71(6): 799-804	Surgery: translabyrinthine approach outcomes	100	Complete removal in 97% of cases, facial nerve anatomically intact in 82%	III
Lunsford et al. Otolaryngol Clin North Am 1992;25(2):471-91	Radiosurgery: outcomes	102	96% tumour control (1,7 years mean follow-up). It is an additional weapon and not replacement to surgery	IV
Cerullo et al. Surg Neurol 1993; 39(6): 485-93	Surgery: outcomes	102	Facial nerve function preserved in 86%, recurrence rate 10%	III
Robinette et al. Am J Otol 1997; 18(6): 378-45	Surgery: hearing preservation	104	Preoperative word recognition is significant predictor	III
Nutik SL. Surg Neurol 1994; 41(1): 28-33	Surgery: facial nerve outcomes	108	Size, experience and ease of dissection important factors	III
Nissan et al. Ear Nose Throat J 1997; 76(1): 37-40	Surgery: facial nerve outcomes	111	Size and approach not significant	III
Nissan et al. Laryngoscope 1997; 107(1): 118-21	Surgery: KTP-532 laser	111	KTR-532 laser is safe and has specific advantages	III
Fucci et al. Am J Otol 1999; 20(4): 495-9	No treatment – radiologic follow-up	119	Most neuromas with periodic MRI do not grow	III
Charabi et al. Otolaryngol Head Neck Surg 1995;113(1):5-14	No treatment – radiologic follow-up	123	Mean follow-up of 3.4 years, 18% of cases showed no growth; 8% smaller	III
Thomsen et al. Tokai J Exp Clin Med 1994; 19(3-6): 93-101	Surgery: algorithm	127	Management algorithm and 74% of neuromas grew with a mean of 3.4 mm/year and 75% lost	III

			hearing preservation candidature while waiting	
Lalwani et al. Am J Otol 1995; 16(6): 758-64	Surgery: delayed facial nerve worsening	129	29% of patients experience delayed facial nerve worsening but with excellent prognosis	III
Lalwani et al. Otolaryngol Head Neck Surg 1994;111(5): 561-70	Surgery: facial nerve outcomes	129	90% of patients H-B scale I or II and the size of neuroma is important	III
Baguley et al. J Laryngol Otol 1992; 106(4): 329-31	Surgery: tinnitus	129	Post-operative tinnitus does not have a significant impact	III
Chung et al. Am J Otol 1997; 18(4): 436-43.	Surgery: quality of life	130	Surgery has not a considerable impact on patients' life	III
Rigby et al. Am J Otol 1997; 18(4): 427-35	Surgery: quality of life	130	Most symptoms are attributable to neuroma and not surgery	III
Van Leeuwen et al. Ann Otol Rhinol Laryngol 1996; 105(6): 423-30	Surgery: quality of life	134	Approach and size correlate with the outcome	III
Andersson et al. Am J Otol 1997;18(4):421-6	Surgery: quality of life	141	Few patients experience social problems post- operatively	III
Andersson G. J Psychosom Res 1999; 46(3):257-60	Surgery: quality of life	141	Anxiety, age and facial nerve function were associated with symptoms	III
Van Roijen et al. Acta Neurochir (Wien) 1997; 139 (1):942-8.	Surgery versus radiosurgery	145	Radiosurgery more cost- effective	IV
Slattery et al. Am J Otol 1997;18(5):596-601	Surgery: middle fossa approach	151	Minimal complications-good to excellent results	III
Dutton, Ramsden et al. J Laryngol Otol 1991;105(3): 165-173.	Surgery: outcomes	151	Mortality 3%, good or normal facial nerve 79%. Size and experience significant.	III
Henrich et al. Ear Nose Throat J 1995;74(7):462-6	Surgery: tinnitus	160	75% of patients report post-operative tinnitus	III
Cohen et al. Am J Otol 1993;14(5):423-33	Surgery: hearing preservation	161	Complications were somewhat increased by attempted such surgery	III
Glasscock et al. J Neurosurg 1993; 78(6): 864-70	Surgery: hearing preservation	161	Hearing preservation achievable in 35% of cases	III

Kondziolka et al. N Engl J Med 1998; 339(20): 1426-33	Radiosurgery: long term outcomes	162	Radiosurgery provides long term control with minimal complications	III
Arriaga et al. Am J Otol 1994;15(5):620-4	Surgery: comparison of approaches	164	Translab and middle fossa similar facial nerve outcomes for neuromas < 1.5 cm	III
Briggs et al. Neurosurgery 1994;34(5):785-90	Surgery: translab removal of large neuromas	167	Acceptable facial nerve function in 75%, complications 4-10%	III
Gormley et al. Neurosurgery 1997;41(1): 50-8	Surgery: outcomes	179	Surgery is preferred over radiosurgery	III
Gormley et al. Neurosurgery 1997; 41(1): 50-60.	Surgery: outcomes	179	Surgery is preferred over radiosurgery in healthy patient	III
Falcioni et al. Am J Otol 1999; 20(5): 660-6	Surgery: complications	200	With proper technique 0% of CSF leak	III
Driscoll et al. Am J Otol 19(4): 491-5	Surgery: complications	210	31% of patients experience post-operative disequilibrium	III
El-Kashlan et al. Am J Otol 1998; 19(1): 104-11	Surgery: vestibular symptoms	220	Significant correlations between pre-op symptoms and vestibular test results and post-op dizziness disability	III
Lynn et al. Am J Otol 1999; 20(4): 484-94	Surgery: complications	237	65% of patients have disequilibrium but with low impact on their life	III
Noren et al. Acta Neurochir Suppl (Wien) 1993;58:104-7	Radiosurgery: outcomes	254	Tumour control in 88%, hearing preservation 77%, trigeminal neuropathy 19%	III
Irving et al. Laryngoscope 1995; 105(8pt1): 809-13	Surgery: complications	257	Nervus intermedius abnormalities are common	III
Irving et al. Am J Otol 1995; 16(3): 331-7	Surgery: quality of life	257	Excellent post-operative quality of life	III
Mass et al. Arch Otolaryngol Head Neck Surg 1999; 125:801-4	Surgery: complications of translabrynthine approach	258	Safe and effective approach	III
Grey et al. Clin Otolaryngol 1996; 21(5): 409-413	Surgery: translabrynthine versus retrosigmoid	276	Retrosigmoid give better facial nerve outcomes	III
Paring et al. Arch Otolaryngol Head Neck Surg 1992; 118(10): 1061-4	Surgery: quality of life	293	Deafness, balance and facial nerve cause the most severe problems post-operatively	III
Symon et al. Br J Neurosurg 1989; 3(3): 343-7	Surgery: outcomes	392	Good or excellent result in 94%, mortality rate 1.4%,	III
Koos et al. J Neurosurg 1998; 88(3): 506-12	Surgery: hearing preservation	442	Hearing preservation from 57% to 100% of patients according to size and location	III
Niranjan et al. Neurosurg Clin	Radiosurgery: hearing	487	21 patients out of 487	III

N Am 1999;10(2):305-15	improvement		experienced hearing improvement	
Niranjan et al. Neurosurg Clin N Am 199; 10(2): 305-15	Radiosurgery: hearing improvement	487	In 4.3% of cases hearing improved	III
Arriaga et al. Otolaryngol Head Neck Surg 1993; 108(3): 220-4	Surgery: facial nerve outcomes	515	Acceptable facial nerve function over 90% of cases	III

Wiegand et al. Laryngoscope 1989; 99(2): 179-87	Surgery: quality of life	541	64% of respondents had some facial weakness and there is a great need for information or patient orientated care	III
Selesnick et al. Am J Otol 1998; 19(6): 846-9	No treatment – radiologic follow-up (meta-analysis)	571	Mean follow-up of 3 years, 46% of cases showed no growth	III
Guerin et al. Ann Acad Med Singapore 1999; 28(3): 402-8	Surgery: complications	611	Surgery is the best treatment for the majority of cases	III
Sampath et al. J Neurosurg 1997; 87(1): 60-6	Surgery: facial nerve outcomes	611	90% of cases House-Brackmann 1 or 2	III
Samii et al. Adv Tech Stand Neurosurg 1995; 22:343-73	Surgery: hearing preservation	653	Hearing preservation rate was 51% in small neuromas and 22% in large neuromas	III
Samii et al. Neurosurgery 1997; 40(1): 11-21.	Surgery: complications	1000	Sub occipital approach and careful selection should reduce mortality rate to <1%	III
Samii et al. Neurosurgery 1997; 40(4): 684-94	Surgery: facial nerve outcomes	1000	Anatomical preservation of facial nerve in 93%	III
Samii et al. Neurosurgery 1997; 40(2): 248-60	Surgery: hearing preservation rate	1000	Hearing preservation is achievable (40% of cases)	III

8.4 The Evidence Base – Conclusions

- 8.4.1 A number of neuromas involute or do not exhibit further growth after initial diagnosis. Thus, the available evidence supports the strategy of watchful waiting with interval scanning in selected cases. No predictors exist that can consistently identify those neuromas that will subsequently increase in size.
- 8.4.2 Evidence demonstrates that surgery can achieve total neuroma removal in up to 97% of patients with mortality, in centres reporting results, of approximately 1%.
- 8.3.4 Some degree of facial paralysis complicates neuroma removal in a significant number of patients and represents a major source of disability.
- 8.3.5 Radiotherapeutic approaches can achieve a ‘control’ rate in a significant number of patients, at least in the short term. Long-term risks, including malignant change and injury to adjacent neural structures, remain a concern. Life long surveillance is needed following treatment.
- 8.3.7 Almost all the above studies assessed each treatment modality separately, were generally retrospective, and thus represent a less than compelling level of evidence (Level III or IV in Table 1).

- 8.3.8 No study exists that systematically compares the different modalities of management (surgery, radiosurgery, interval scanning).

9. Quality of Life Following Acoustic Neuroma Surgery

- 9.2 Quality of life studies comparing post-operative quality of life with pre-operative status need to be interpreted with caution. When most patients present with an acoustic neuroma, they enjoy excellent health. They thus opt for surgery with the aim of preserving life and preventing major neurological complications, possibly many years later.
- 9.2 Another important shortcoming in assessing quality of life and comparing the different modalities of treatment is the lack of standardised and validated instruments that can reliably measure and compare the quality of life in patients with acoustic neuroma across the various methods of management.
- 9.3 Using a validated health-status instrument (The Glasgow Benefit Inventory), it was found that 54% of UK patients experienced a poorer quality of life after surgery as compared to their pre-operative status (Nikolopoulos et al 1998). Fifty per cent engaged in fewer social activities after surgery. Surgery had a greater impact on the life quality of younger patients. A statistically significant correlation between quality of life and neuroma size was not found. These results are similar to those reported by a Danish Study (Jorgensen and Petersen 1994) but at variance with a study by Irving et al (1995) and Chung et al (1997) who found surgery had minimal impact on life quality.
- 9.6 A further UK study (Bateman et al 2000), using open-ended questionnaire techniques, categorised patients' symptoms according to the World Health Organisations classifications of impairment, disability and handicap. The study revealed a wide variety of post-operative symptoms, especially visual disturbance (49% of patients) and psychosocial deficits (34% of patients)
- 9.7 A Dutch study found 25% of surgical patients were declared unfit for work following surgery compared with an American study showing that only 1.6% became unemployed after surgery (Van Leeuwen et al 1995 & 1996, Chung et al 1997). However it is clear that the recovery time following stereotactic radiosurgery is shorter than that after microsurgery. Those patients in the surgical group with paid jobs were absent from work for 3 months on average. Patients with jobs undergoing stereotactic radiosurgery were absent for one working day (Van Roijen et al 1997).
- 9.6 One UK study of a consecutive series of surgically treated patients found that almost 80% of patients continued without change with their usual occupations (Nikolopoulos et al 1998).
- 9.7 Patients and surgeons do not always agree about outcomes, especially concerning facial nerve function following surgery (Wiegand et al 1989). Nevertheless, an objective independent assessment of facial nerve function, for example using the House Brackmann scale, can be a useful index of this specific neurological limitation following different methods of management.
- 9.8 Cross et al (2000), using 4 validated questionnaires, found that the distress experienced by patients with facial paralysis following acoustic neuroma surgery does not correlate with the grade of the paralysis as attributed by surgeons. They found that even patients with minimal disturbance of facial nerve function may experience great personal distress (Cross et al 2000).

10. Cost - Effectiveness

- 10.1 No formal studies exist that compare the cost-effectiveness of the various treatment options in acoustic neuroma patients.
- 10.2 One Dutch study compared the costs of microsurgery with those of stereotactic radiosurgery (Van Roijen et al 1997). This demonstrated that the direct costs of surgery exceeded those of radiosurgery by 20%. However, the outcomes of these two treatment modalities are not equivalent. Surgery results in total neuroma removal whereas radiosurgery does not and requires the patient to undergo life-long serial scanning.

11. Indicators of Good Practice

- 11.1 The care of acoustic neuroma patients requires multidisciplinary teamwork with access to the full range of specialist support services these patients may need.

Centres offering this care should have:

- 11.2 An otolaryngologist and a neurosurgeon with a specialist interest and training in neuro-otology and skull base surgery as well as access to specialist facilities in stereotactic neurosurgery.
- 11.3 There should be links and communication between those neurosurgeons with specialist expertise in stereotactic radiation therapy (at a supraregional or national level) able to contribute both to the clinical management in broad terms as well as recommend and supervise intervention by stereotactic radiation therapy.
- 11.7 Audiological facilities to undertake diagnostic auditory and vestibular investigations as well as post-operative auditory and vestibular rehabilitation. Post-operative dizziness and tinnitus can be troublesome and patients may need supportive therapy. The provision of CROS hearing aids should be considered in appropriate patients. Access to a speech and language therapist specialising in swallowing difficulties may at times be necessary. Referral to an audiological physician may be desirable in the rehabilitation of certain patients.
- 11.8 Neurodiagnostic imaging and neuroradiological support, including facilities for emergency imaging.
- 11.9 Neuroanaesthetic provision consistent with standards described in 'Guidance on the Provision of NeuroAnaesthesia' (pages 60-65) in Guidelines for the provision of Anaesthetic Services published by the Royal College of Anaesthetists - July 1999.
- 11.7 Neuromonitoring facilities for monitoring the facial nerve and, where indicated, hearing function during surgery.
- 11.8 Access to intensive care facilities.

- 11.9 Patient numbers: the centre should have sufficient level of clinical activity to gain familiarity with the diverse needs of acoustic neuroma patients as well as to develop and maintain surgical skills and provide surgical training.
- 11.10 Audit – regular multidisciplinary audit of treatment outcomes.
- 11.11 Plastic and Reconstructive Surgery: access to these facilities, especially for facial reanimation surgery. Access to oculoplastic surgical expertise may be helpful in the management of eye-lid dysfunction.
- 11.12 Referral to voluntary support agencies when requested.

12. The Patients' Perspective

- 12.1 A number of patient organisations, such as the British Acoustic Neuroma Association, offer a network of support for patients. Patients should be made aware of the existence of these organisations and should be helped to contact them if they so chose.
- 12.2 The RNID is the largest charity representing the 8.7 million deaf and hard of hearing people in the UK. It supports initiatives which provide evidence-based information to empower people to make informed choices. The RNID therefore welcomes the production of these guidelines as a means of assisting professionals and patients in decisions relating to the diagnosis and management of acoustic tumours to improve the quality and consistency of patient care.

13. Audit and Clinical Governance

- 13.1 A national audit of acoustic neuroma treatment results has not been undertaken.
- 13.2 A national audit would enable units to compare their performance across patient groups and would help the implementation of clinical governance in acoustic neuroma practice. However, validated clinical outcomes need to be developed to ensure meaningful comparisons are made across treatment modalities. It would also facilitate the accrual of sufficient numbers to compare treatments in a prospective manner with sufficient statistical power.
- 13.4 With agreement between teams on a minimum data set and with appropriate administrative support, such studies could be readily undertaken.
- 13.4 Centres offering SR and FSR should be limited in number and identified nationally. Appropriate criteria and a process of designation need to be developed. They should offer life-long surveillance and make available their results for treating acoustic neuroma patients, especially their long-term outcomes.

14. Equity of Access

- 14.1 All patients with acoustic neuromas should have access to a uniformly high standard of care. Evidence suggests that patients achieve the best outcomes in centres with a special interest in this condition. Therefore, patients should be referred to a specialist unit known to have the expertise in acoustic neuroma management (which may not be the nearest hospital).

15. Health Promotion

- 15.1 Greater public awareness of the importance of unilateral or asymmetrical auditory symptoms would do much to facilitate earlier diagnosis of acoustic neuromas. *'Its time to test your hearing'* by the Royal National Institute for the Deaf People (RNID) is an example of a programme which aims to educate the public about hearing disorders. The RNID has also produced a factsheet on acoustic neuroma.
- 15.2 General Practitioners (GP's). The first point of contact with the health care system for most patients remains the family practitioner. Continuing Education programmes should remind GP's of the importance of referring patients who present with unilateral or asymmetrical auditory symptoms to their local ENT Department. The introduction of hearing testing into GP Health Screening would be particularly helpful.
- 15.3 When patients are referred directly to audiology departments, protocols for onward referral of those patients with findings suggestive of acoustic neuromas should be implemented.

16. Updating

These guidelines will be updated in March 2003.

17. Reference List

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