Arq Neuropsiquiatr 1997;55(3-A):393-402

ACOUSTIC NEURINOMAS

DIAGNOSIS AND TREATMENT

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ABSTRACT - We present our experience with 83 patients with acoustic neurinomas (January 1988 to November 1996); 81 patients underwent surgery and 2 patients with intracanalicular neurinomas received conservative therapy due to their advanced age (1 case) and patient's option (1 case). The surgical approach was retrosigmoid/ trasmeatal and the goal was total removal in one procedure with preservation of facial and cochlear nerves. Radical removal of lesion was attempted in 79 cases and it was possible in 77 patients. Subtotal tumor resection was planned in 2 cases. Facial nerve function (grades I to III, House and Brackman) was preseved in 90% and hearing in 58% of those with preoperative hearing. Three patients died due to postoperative complications. Early diagnosis of acoustic neurinomas is the most important factor in the prognosis and is one of the most important conquest of neurophysiology and modern neuroimaging. The management of these patients still present many controversial points. This article presents the diagnostic procedures used for acoustic neurinomas, the advantages and disadvantages of the different surgical approaches and the alternative management of these lesions.

KEY WORDS: acoustic neurinoma, vestibular schwanoma, facial nerve, hearing preservation, radiosurgery, management.

Neurinoma do acústico: diagnóstico e tratamento

RESUMO - Apresentamos nossa experiência no tratamento de 83 pacientes com neurinomas do acústico (Janeiro-1988 a Novembro-1996). Cirurgia foi realizada em 81 pacientes e tratamento conservador (idade avançada e escolha do paciente) em 2 casos de neurinomas intracanalicures. O objetivo da cirurgia foi ressecção radical com preservação dos nervos facial e coclear. Em 77 de 79 pacientes foi obtida remoção total. Em 2 casos foi proposta ressecção sub-total devido às más condições clínicas (1 paciente) e por tratar-se de neurofibromatose tipo 2 com lesão dos nervos facial e coclear do outro lado (1 paciente). Preservação do nervo facial (grau I a III, House e Brackman) foi possível em 90% dos casos com função facial pre-operatória normal. Dos 12 pacientes que apresentavam audição no pré-operatório, 7 mantiveram a audição. Ocorreram 3 óbitos no pós-operatório imediato. As vantagens e desvantagens dos acessos cirúrgicos mais utilizados na cirurgia do neurinoma do acústico são demonstradas. O fator mais importante no prognóstico desses pacientes é o diagnóstico precoce, possível com o desenvolvimento de novos métodos neurofisiológicos e neurorradiológicos. O tratamento no entanto pode apresentar algumas controvérsias, variando desde o simples acompanhamento clínico à remoção cirúrgica ou o uso de radiocirurgia.

PALAVRAS-CHAVE: neurinoma do acústico, schwanoma vestibular, nervo facial, preservação da audição, radiocirurgia.

At the beginning of this century surgical treatment for acoustic neurinomas carried a very high and prohibitive mortality[4,5,6]. Subtotal removal was proposed by Cushing[4] in order to achieve

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better results. Dandy[6] published some years later a series of patients with radical removal of acoustic neurinomas with an acceptable mortality for those days. This paper opened, however, the possibility of total removal of the lesion with cure of the patient. Atkinson[1] demonstrated that lesion of the anterior inferior cerebellar artery (AICA) was the main cause of mortality. Olivecrona[27] published a large series with low mortality and preservation of facial nerve. The introduction of the surgical microscope opened a new horizon in the surgery of these lesions. The pioneering works of House[12] and Rand & Kurze[31] began a new era in the management of acoustic neurinomas. Radical removal and preservation of the facial nerve became a routine procedure. In the late 80s with the development of new diagnostic tools such as high definition CT-scan, brain stem evoked potentials and magnetic ressonance imaging (MRI), the diagnosis of small tumors with serviceable hearing became possible[14,17,36]. Preservation of choclear nerve and hearing function was the next goal in the development of this surgery. Protocols of radiosurgical treatment have been developed with promising results, but long-term follow-up from multiple centers will be required for better evaluation of this method.

The new diagnostic and therapeutic procedures have allowed a much better follow-up and understanding of the natural history of these lesions. Size of lesion, clinical condition of patient, preoperative hearing function, experience of surgeon and wish of patient are the most important factors involved in the management decision. Choice of surgical approach is also very important. The three main surgical approaches used are: retrosigmoid-transmeatal, translabyrinthine and middle fossa. Each of these approaches has advantages and disadvantages. In our experience the retrosigmoid-transmeatal approach gives the best surgical exposure of the lesion, control of facial nerve inside of the internal auditory canal and at brain stem. Moreover it gives the lowest rate of postoperative complication and the possibility of hearing preservation.

In this article, we report our experience with the management of a series of 83 patients with acoustic neurinomas. The results concerning radicality of removal, preservation of facial and coclear nerves and mortality are presented. We also assessed the timing of surgical indication and advantages and disadvantages of the three different surgical approaches.

MATERIAL AND METHODS

During a 9-year period (January 1988 - November 1996) 83 patients with acoustic neurinomas were treated in our clinic. The age of the patients varied from 16 to 78 years of age. Tumor size was larger than 3.0 cm in 65 (78%) and smaller in 18 (22%) of the cases (Figs 1 and 2). Ten patients suffered previous operations in other clinics with partial tumor removal and complete facial nerve lesion in 5, two of these received conventional radiation therapy before admission to our clinic. Twelve patients presented some hearing preoperatively. Two presented hearing classified as "non-serviceable" with scores under 50 db of hearing and 50% speech discrimination. Six patients had neurofibromatosis (NF 2) with bilateral acoustic neurofibromas and other tumors including cranial nerves neurofibromas, spinal neurofibromas, meningiomas and astrocytomas.

Eighty-one patients were operated on. Two additional cases were treated conservatively. In the first one, the tumor was very small (5 mm) and asymptomatic and the patient declined surgery. The second patient was 75-year-old with a 15 mm typical acoustic neurinoma in the MRI. This patient was already deaf and the tumor did not grow in a follow-up MRI 6 months after the diagnosis was made. The retrosigmoid-transmeatal approach was used in 80 cases. In one case with NF 2, bilateral surgical removal was performed. The last patient (78-year-old, in poor clinical condition) with a 40 mm cystic acoustic neurinoma underwent sterotactic biopsy and aspiration of the cyst.

Forty-eight tumors were approached in dorsal surgical position ("mastoid position") with the head rotated to the opposite site. The semi-sitting position was used in 20 cases and the lateral position in 13 (Fig 3). The choice of surgical position was based on the size of tumor and presence of other lesions such as high cervical neurofibromas and astrocytomas.

Intraoperative monitoring of facial nerve was performed in 40 patients and intraoperative acoustic brainstem evoked potentials (BAEP) were recorded in 10 cases with an Amplaid Multisensorial Monitoring System (Milan,

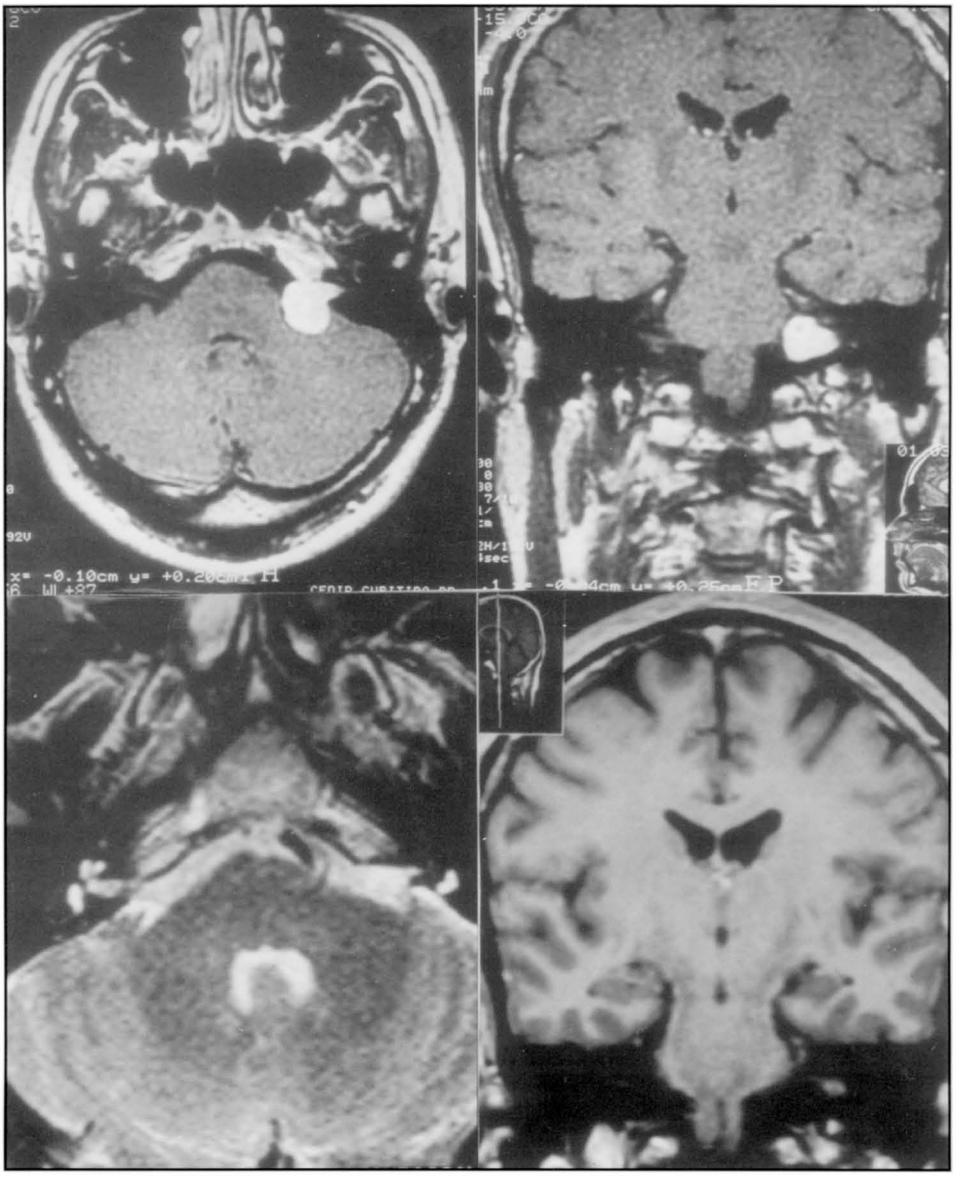


Fig 1. Upper: Preoperative MRI examinations (T1 with gadolinium) showing an acoustic neurinoma with 2.0 cm. Below: Postoperative MRI examinations (left T2 and left T1 with gadolinium) after total removal of the lesion with preservation of cranial nerves VII and VIII. The facial and cochlear function could be preserved in this case.

Italy). The muscle electrodes used to record intraoperative responses from *orbicularis oris* muscle were applied at the nasolabial groove after the patient has been anesthetized but before surgery has begun. The anesthesiologist used muscle relaxants only at the beginning of operation in order to allow monitoring of the EMGs related to facial nerve. The facial nerve was stimulated with monopolar electrodes with impulses up to 1 mA which were recorded during 20 microseconds or with direct manipulation or traction on the facial nerve. This technique alerts the surgeon to the proximity of the nerve; a loudspeaker provided an acoustic feedback. The cochlear nerve was monitorized by recording brain stem auditory evoked potentials (BAEP). Following final patient positioning,

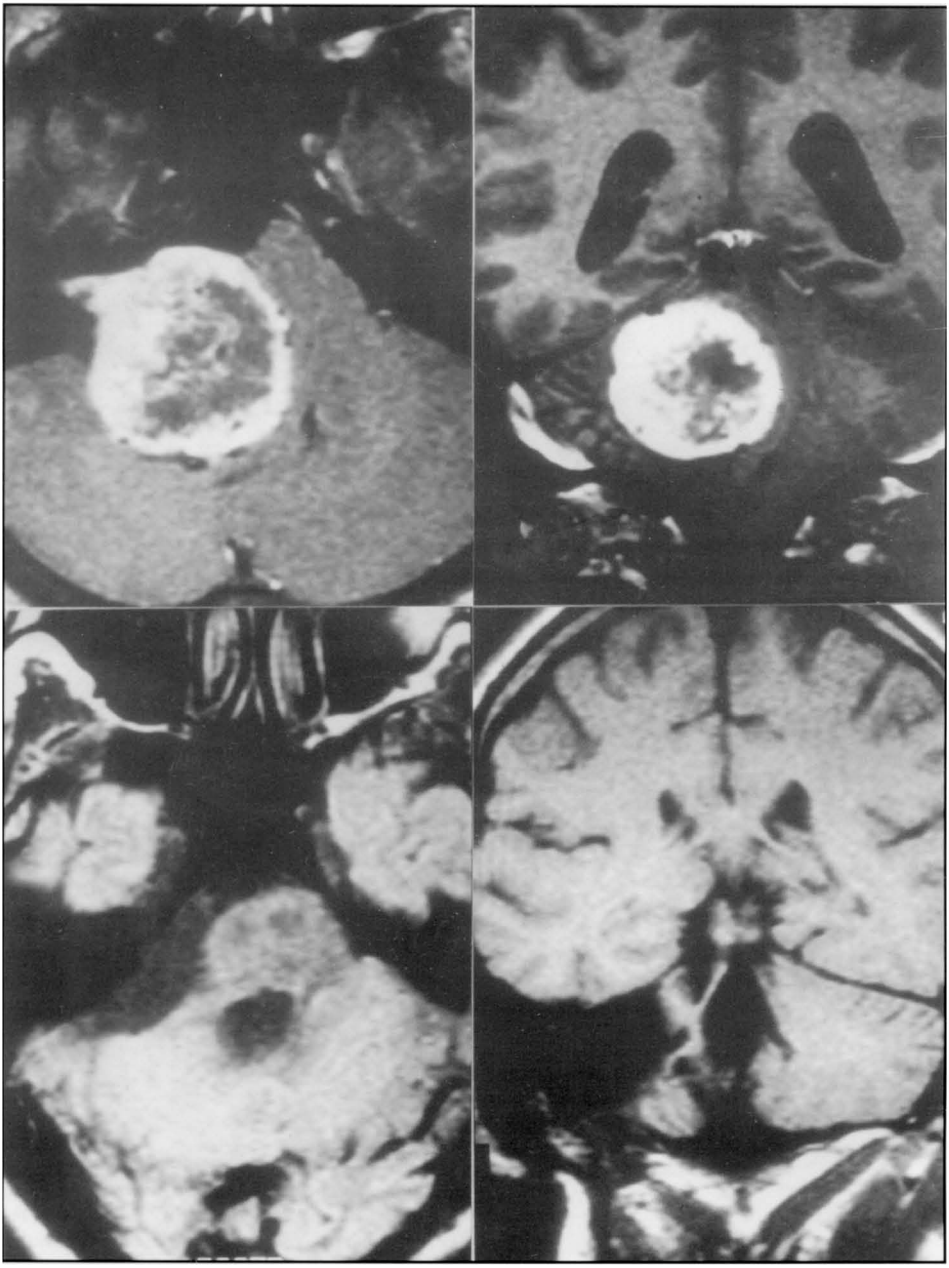


Fig 2. Upper: Preoperative MRI examinations (T1 with gadolinium) of a patient with a large acoustic neurinoma (4.5 cm) compressing the brain stem and the IV ventricle. Below: postoperative MRI (T1 with gadolinium) after total removal of the lesion and preservation of the facial nerve.

the vertex (Cz) and ear recording electrodes were placed. The ear electrodes are placed in the pinna of the earlobe for both sides. The sound stimuli were clicks presented through miniature stereo earphones at an intensity of 100 dB nHL which are recorded during 10 ms. The earphones were sealed in the outer ear with adhesive tape and the first five peaks of BAEP (waves I through V) are recorded continuously.

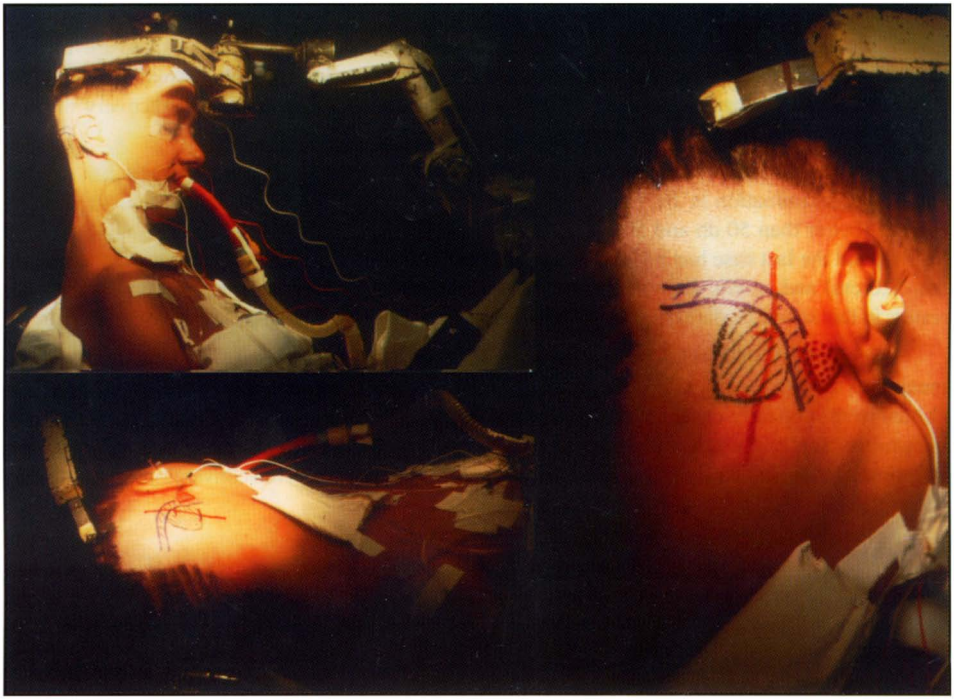


Fig 3. The surgical position used: Left upper: the semi-sitting position, Left below: the lateral (mastoid) position and right the surgical incision 2.0 cm behind the mastoid tip. Electrodes for monitorization of the facial and cochlear nerves are placed.

All patients had postoperative CT-scan or MRI control examinations and the follow-up period was 4.5 ± 3.0 years (mean ± sd).

RESULTS

Total removal of the lesion was achieved in 77 out of 80 microsurgical treated patients. In two patients subtotal removal was done due to extremely high tumor vascularization in one case and adhesion to the brain stem in a patient with a 55 mm tumor. These two patients were followed 20 and 28 months respectively and the small tumor remnant showed no size increase in this period of time. In one NF 2 patient subtotal removal of a 45 mm tumor was performed because he presented a facial palsy of the opposite side. Stereotatic cyst aspiration and biopsy was indicated in one patient due to poor clinical condition. This patient remained asymptomatic after the procedure and a MRI-examination 6 months after surgery revealed complete resolution of the cyst and a small (15 mm) tumor remnant in the cerebellopontine angle and internal auditory canal. One patient with a 50 mm cystic acoustic neurinoma presented a tumor recurrence 2 years after a "total microsurgical removal". This patient remained asymptomatic and a new surgical removal was proposed and refused.

The facial nerve function (grades I to III of the House and Brackmann classification[13]) could be preserved in 69 patients (90%) of 76 patients with normal preoperative facial nerve function. Facial nerve function was preserved in all patients with tumor size smaller than 2.5 cm. Five patients had preoperative facial nerve palsy due to surgery in other clinics. Patients with anatomically preserved facial nerve during surgery and with postoperative facial weakness were followed for at least one year. All of them recoreved to grades II or III. Patients with intraoperative facial nerve lesion underwent surgical reconstruction of the facial nerve. In one patient, an end to end anastomosis was carried out at the time of tumor removal and he had a grade II recovery. In the remaining 16 patients

with facial nerve lesion (5 preoperative and 11 postoperative), XII-VII anastomosis was performed 2 weeks after tumor removal. Recovery to grades III and IV was obtained in all cases. In three patients, a modification of the classical technique was used. Only a half part of the splitted hypoglossal nerve and the *ansa hypoglossi* were sutured to the distal part of facial nerve. Recovery of these patients was grade III and minimal atrophy of the tongue was observed.

Twelve patients presented preoperatively with some hearing. Hearing was better than 50 db and 50% discrimination in 10. Preservation was possible in 7 cases, 5 remained with serviceable hearing (better than 50 db and 50% discrimination), one developed postoperative non-serviceable hearing and the last case had pre and postoperative non-serviceable hearing. Tumor size (6 smaller than 2.0 cm) and careful intrameatal microsurgical dissection were the main reason for the good results in this series. No late deterioration of hearing was observed in the follow-up of these patients.

Surgical complications were cerebrospinal fluid (CSF) rhinorrea in 7 patients (9%) with meningitis in 4 (5%) and CSF leak through the wound in 2, hematoma in tumor bed in 3 cases (which were surgically evacuated). Two patient died (2,5%) after the surgery due to intracerebellar hematomas and brainstem infarction. One patient died 12 days after surgery due to pulmonary infection. This patient had been operated before in other clinic and received conventional radiotherapy.

DISCUSSION

The first point to be discussed is the terminology of these lesions[34]. These tumors are usually called acoustic neurinomas but the preferred name should be vestibular schwannoma, because it is composed of Schwann cells and most frequently arises from the vestibular portion of the VIII cranial nerve. These lesions are usually unilateral, with onset in middle age. Bilateral lesions are associated to neurofibromatosis (NF 2) and frequently other lesions are observed. Unilateral hearing loss with or without tinnitus is the most frequent symptom of these patients. The neuro-otologic diagnosis is completed with puretone air and bone conduction thresholds, speech reception thresholds, speech discrimination scores and brain stem auditory evoked response. Neuroradiological examination involving gadolinium enhanced MRI usually confirms the diagnosis even in case of small lesions. High definition CT-scan with bone window should always be performed in order to identify semicircular canals and jugular bulb.

The management of acoustic neurinomas has changed considerably with the development of new diagnostic possibilities and microsurgical procedures. The first successful removal of such a lesion is credited to Sir Charles Ballance in 1894. The patient survived after an unsterile tumor removal with blunt finger dissection[2]. At the beginning of this century the mortality for surgical removal of an acoustic neurinoma was unacceptably high (over 80%). This fact led Cushing to propose intracapsular-subtotal removal of the lesion with a mortality of 11%[4]. Dandy[5,6] in 1941 published a series of patients with total removal of the tumors and a mortality rate of approximately 2.4%. Facial nerve paralysis, deafness associated to cranial nerves V, IX, X and XI paresis were usually the consequences of this surgery. In 1964, the otologist William House[12] from Los Angeles began a new era in the treatment of these lesions using microsurgical techniques and reported a series of patients with preservation of the facial nerve in 95% of the cases and a mortality of 5.4%. In 1965 Rand and Kurze[32] neurosurgeons from the same city published their experience using a retrosigmoid- transmeatal approach. Since these early microsurgical times an increasing number of series with radical removal of the lesion and preservation of the facial nerve function and hearing with almost no mortality have been published[17,22,24,26,30,32,35,42]. In the last years series of patients treated with radiosurgery have been studied but long term follow-up is necessary to evaluate the indications and results of this method[18,19,23,24,29]. The natural history of these lesions is not well known. Most papers show a slow grow with an increase in size, which varies from 0 to 10 mm per year; however, a longer follow-up of these patients is necessary[39,41]. Molecular genetic analysis demonstrated chromosome 22 allele loss in patients with sporadic (non NF2) acoustic neurinomas[15].

The optimal strategy for managing these patients depends on several factors as: size of the lesion, age and general condition of the patient, experience of the surgical team, availability of sophisticated technique as intraoperative monitoring of facial and cochlear nerve function and radiosurgery. Options for the management include observation, surgery and stereotactic radiosurgery[7,11,26,28,29,34,36,37,42]. In our opinion conservative treatment should be reserved for elderly patients with small asymptomatic lesions or younger patients, who are already deaf, with small lesions (intracanalicular). If the patient has good hearing and a small intracanalicular tumor we indicate surgery as the best possibility for preservation of cochlear nerve function. Age is usually not the most important factor, the general condition of the patient should be taken in consideration for indication of surgery. The goals of surgical treatment are: total removal of the lesion, preservation of facial nerve and if possible the function of cochlear nerve. Radical removal of the tumor is possible to be achieved in almost all patients and preservation of facial and coclhear nerve function is higly dependent of tumor size.

Three main surgical approaches (middle fossa, translabyrinthine and retrosigmoid) have been used with excellent results[3,9,12,16,25,33,38,42]. The middle fossa approach is used to expose only small tumors within the internal auditory canal and hearing may be preserved with this surgery. Extradural retraction of the temporal lobe and manipulation of facial nerve inside of the internal auditory canal (IAC) is needed to expose the lesion. The translabyrinthine approach has the main disadvantage of sacrificing hearing and may not expose adequately the brainstem and posterior fossa mainly in cases of very large lesions. Identification and protection of the facial nerve early in the course of the procedure is the main advantage of this procedure. According to our experience the retrosigmoid-transmeatal approach permits adequate surgical exposition of all tumor sizes, preservation of hearing, identification of facial nerve inside of IAC and control of vessels and nerves of the cerebellopontine angle. Cerebellar retraction and inadequate exposure of the IAC fundus has been reported as the main disadvantage of this procedure. Using correct technique with removal of the retromastoid air cells, exposition of sigmoid sinus and opening of the lateral aspect of the *cisterna magna*, almost no retraction of the cerebellum is needed. The posterior and lateral portions of the IAC are opened with diamond drill exposing the fundus, allowing identification of the vestibular, facial and cochlear nerves. No patient in our series presented postoperative cerebellar edema. The retromastoid-transmeatal approach has been used for all acoustic neurinomas in our clinic (Figs 4 and 5).

Preservation of facial nerve is associated to the size of lesion. The overall preservation rate for all tumor sizes varies from 54% to 89%[17,26,34,42]. In our cases an overall preservation of 90% could be achieved and in all patients with tumor smaller than 2.5 cm. Preservation of hearing depends on size, surgical approach and experience of the surgeon[16,17,21,25,36]. In a review of the English literature Gardner and Robertson[9] found 33% of hearing preservation in 621 patients. In our series from 12 patients with preoperative hearing preservation was possible in 7. These results were obtained due to three main factors: choice of surgical approach, size of lesion (6 smaller than 2.0 cm) and careful surgical dissection of the cochlear nerve within the internal auditory canal. In one case the pre and postoperative hearing was considered "non serviceable". According to our philosophy hearing should always be preserved because even if the postoperative hearing is classified as "not useful" it is much superior to that achieved with cochlear implant technique for deafness. Additionally in the future these patients may have benefits from new techniques if they remained with some cochlear nerve function. Postoperative tinnitus may be a complication of cochlear nerve preservation[10]. It was not observed in our cases.

Intraoperative monitorization of facial and cochlear nerves may be useful to achieve better results with the preservation of these nerves. Direct stimulation of facial nerve with monopolar electrode or with mechanical stimuli facilitates identification of facial nerve during tumor dissection. BAEP are recorded to know the integrity of cochlear nerve function. This technique has some disadvantages: BAEP have a small amplitude in relation to the ongoing electrical activity of the

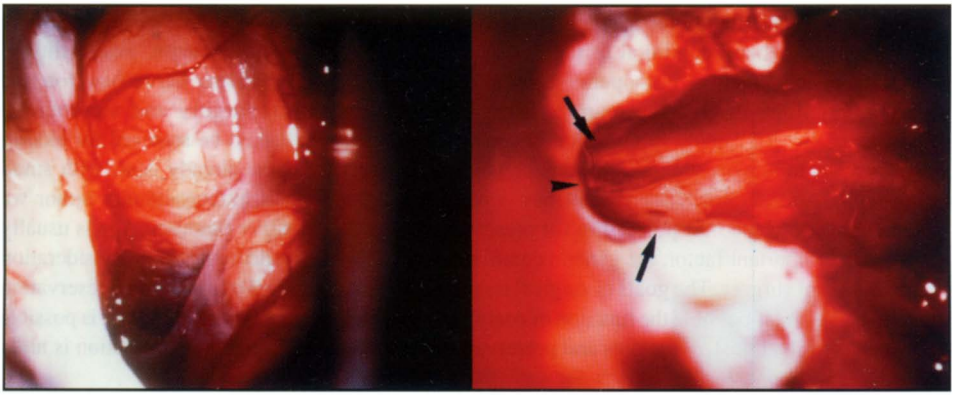


Fig 4. Intraoperative picture showing a large left side acoustic neurinoma (left). After total removal the facial and cochlear nerves (arrows) can be seen inside of the IAC (right). The transverse crest is identified at the fundus (arrow head).

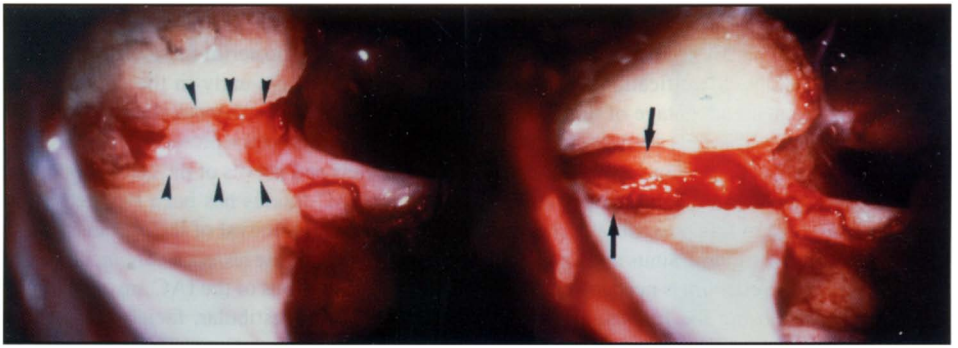


Fig 5. Intraoperative picture showing an intracanalicular neurinoma (arrow heads) and the VII & VIII nerve bundle (left). The tumor was totally removed and the facial and cochlear nerves preserved (arrows). The vessels of VII & VIII cranial nerves could be preserved (right).

brain and averaged responses must be obtained for few minutes to give an interpretable record. This makes real time recording impossible. This technical limitation could be circumvent through direct recording of cochlear nerve action potentials in the cochlea.

The incidence of CSF-leak through the incision or rhinorrhea is almost the same with translabyrinthine or retrosigmoid-transmeatal approaches and varies from 4 to 10%[20,26,34,40]. Meningitis occurs in about 3% of the cases[20]. In our cases we had 9% of CSF-leak with 5% meningitis. Radical removal of the lesion is the main goal of the surgery. It could be obtained in 96% of our cases. In 2 occasions radical removal was not possible because of very high vascularization of tumor and tight adhesion to brainstem in patients with very large lesions. One 78-year-old patient in poor clinical condition underwent stereotactic aspiration of a large cystic acoustic neurinoma and biopsy. These three patients remained asymptomatic postoperatively and the tumor remnant did not grow in postoperative MRI control examinations 28, 20 and 5 months respectively. One patient presented tumor recurrence 2 years after "radical resection" of a large cystic lesion. He remained asymptomatic and refused a new surgical procedure. Two patients died in this series due to postoperative hemorrhage in the tumor bed. These patients had large lesions and in spite of early recognition and drainage of the hematomas died of brainstem infarction. Another patient died 12 days after surgery due to pulmonary infection. This patient had been operated before in another institution with sub-total

removal and was submitted to conventional radiotherapy. Total removal was possible but she developed swalling disturbances with aspiration pneumonia.

Stereotactic radiosurgery has been used as primary therapy or after subtotal removal of the lesion but the results should be analyzed carefully[24,29]. The complications after the earlier Swedish series was unacceptably high and the new series of patients treated with lower dosis need a follow-up of a decade or longer to better know the results. A short follow-up of 2 to 3 years is a concept for malignant tumors and not applicable to slow growing lesions. The goal of stereotatic radiosurgery is to control tumor size whereas with microsurgery is to completely remove the tumor. In a recent series of patients treated with radiosurgery, 60% of the cases remained unchanged in size and 6% increased, in only 34% of these patients the lesions reduced in size[29]. In patients with neurofibromatosis radiation therapy may induce the development of neurofibrosarcomas[8].

In conclusion, the diagnosis and surgical treatment of acoustic neurinomas became at the end of this century in specialized clinics a routine and safe procedure.

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