The Laryngoscope

Case Report

Cochlear Schwannoma Removed Through the External Auditory Canal by a Transcanal Exclusive Endoscopic Technique

Livio Presutti, MD; Matteo Alicandri-Ciufelli, MD; Elisa Cigarini, MD; Daniele Marchioni, MD

Surgical approaches to the inner ear and internal auditory canal (IAC) are widely known and extensively recorded. The most popular can be classified as retrosigmoid, transmastoid-translabyrinthine, and middle cranial fossa approaches. For the first time, an exclusive endoscopic approach to the IAC is described here, used to remove a cochlear schwannoma involving both the IAC and labyrinth. The operation provided a direct transcochlear intradural approach from lateral to medial and from external to internal auditory canal, without any external incision. The pathology was totally removed, and the postoperative outcome of the facial nerve was grade II (House-Brackmann grading system) at 3-month follow-up.

Key Words: Inner ear; internal auditory canal; cochlear schwannoma; transcanal approach; endoscopic ear surgery.

***Laryngoscope*, 123:2862–2867, 2013**

INTRODUCTION

Surgical approaches to pathology extending into internal auditory canal (IAC), such as acoustic neuromas (ANs), are widely known and have been extensively recorded. The most popular can be classified as retrosigmoid, transmastoid-translabyrinthine, and middle cranial fossa approaches. Clinical indications, advantages, disadvantages, and risks in terms of mortality and morbidity have been carefully described.[1] A common factor in all of the methods described to date is that all are indirect approaches to the inner ear; the retrosigmoid and translabyrinthine methods approach the pathology posteriorly, while the middle cranial fossa method approaches the pathology superiorly. To access the internal auditory canal (IAC) and cerebellopontine angle (CPA), all of these approaches require wide external incisions and a variable degree of temporal bone removal.

The first introduction of the endoscopic technique in IAC surgery has been in combination with the retrosigmoid approach[2] after removal of the CPA extension of the neoplasm. The intracanalicular extension was removed under endoscopic control, trying to avoid extensive drilling of the posterior aspect of the petrous bone. A gradual introduction of endoscopic techniques to middle ear pathology treatment has taken place since the 1990s.[3] Endoscopy was used primarily for the visualization of hidden areas, such as the posterior epitympanum during classic microscopic tympanoplasties.[4] Gradually it was also used in surgery to replace the microscope as the main tool during middle ear operations.[5–8] At present the main application of endoscopic surgery is in the surgical treatment of middle ear cholesteatoma, but with the natural evolution of the technique there have been advances in lateral skull base surgery. In recent years the authors progressively noticed that the internal ear and all of the temporal bone could also be accessed in an endoscopic-assisted fashion or even with an exclusive endoscopic approach. The problem would only be to record the landmarks and procedures as thoroughly as possible, and to integrate them in classical microscopic approaches.

The development of these endoscopic techniques required several cadaver dissections to better understand the anatomy and to define appropriate instruments for this purpose. During these dissections some advances were made in exploring the internal ear: from the labyrinth, jugular, and carotid to the IAC, until an appropriate procedure was recorded and ready to be applied clinically. For the first time an exclusive endoscopic approach to the IAC is described here, and used to remove a cochlear schwannoma (CS) involving both the IAC and cochlea. The operation used a direct transcochlear approach from lateral to medial and from external to internal auditory canal, without any external incision. The aim of this article is to describe the surgical technique used, and to discuss possible future applications of this approach.

CASE REPORT

In November 2011 a 40-year-old man presented to the Ears, Nose, and Throat (ENT) emergency room with vertigo, tinnitus, and hearing loss in his left ear. During

From the Otolaryngology–Head and Neck Surgery Department, University Hospital of Modena, Via del Pozzo 71, 41100, Modena, Italy

Editor's Note: This Manuscript was accepted for publication February 4, 2013.

The authors have no funding, financial relationships, or conflicts of interest to disclose.

Send correspondence to Elisa Cigarini, MD, Otolaryngology–Head and Neck Surgery Department, University Hospital of Modena, Via del Pozzo 71, 41100 Modena, Italy. E-mail: eliciga@hotmail.com

DOI: 10.1002/lary.24072

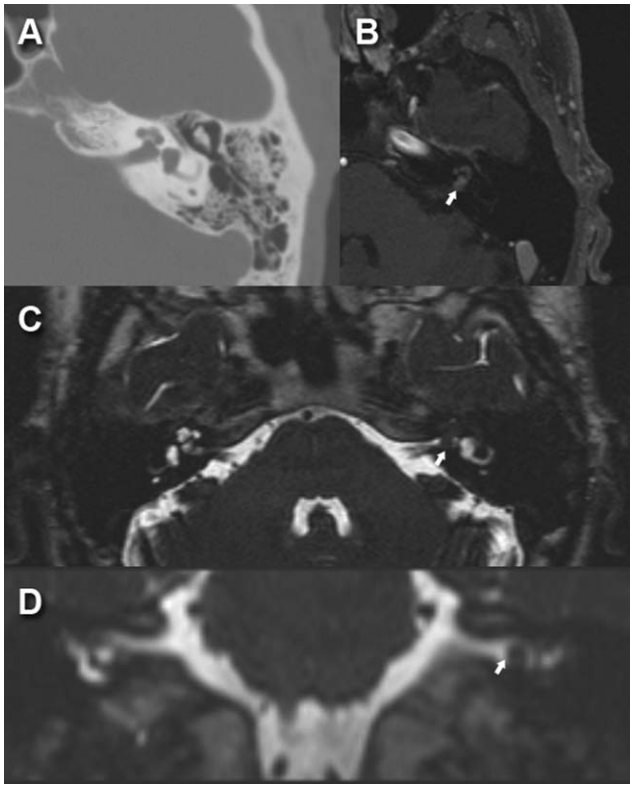

Fig. 1. Preoperative radiologic evaluation. (A) At computed tomography (CT) scan, no obvious erosions involving the inner ear or internal auditory canal were evident. (B) Axial T1-weighted MR images, showing the neoplasm, involving the cochlea and internal auditory canal in its most lateral portion. The mass is hyperintense and is slightly enhanced by gadolinium (arrow). (C) Axial T2-weighted MR images. The mass appears hypointense to muscle (arrow). (D) Coronal T2-weighted MR images. The involvement of the internal auditory canal by the small mass is highlighted (arrow).

the clinical history he referred three emergency evaluations for recurrent dizziness over the last year. Clinically, at the last emergency admission, a second-degree right nystagmus was evident, and he complained of vertigo and tinnitus in the left ear. Clinical manifestations were similar to earlier episodes. Due to the marked neurovegetative symptoms he was admitted to the ENT department inpatient clinic for further evaluation. During his hospital stay an audiometric exam was performed; results were normal for the right ear, whereas for the left ear there was an almost total deafness, with only residual hearing of higher frequencies. The vestibular examination (caloric testing) showed an absence of left vestibular reflexes and a normal vestibular function contralaterally. The brain computed tomography (CT) scan did not show any anomaly (Fig. 1A). At the brain 1·5 T magnetic resonance imaging (MRI) with contrast medium infusion (gadolinium), an isointense mass at the left cochlear membranous labyrinth was seen in T1-weighted images, sparing the apical turn of the cochlea with a portion invading the IAC; the lesion uptook gadolinium (Fig. 1B). On T2-weighted images, the lesion appeared iso/hypointense to the brain (Fig. 1C, D). These findings were compatible with CS with intracranial extension.

During hospitalization the patient followed a therapy based on 20 mg of methylprednisolone 2 times a day, and levosulpiride as needed for nausea. The patient was discharged after 5 days, with prednisone therapy (25 mg per day for 5 days). Vertiginous symptoms at discharge had significantly improved; nystagmus was no longer present, while subjective hearing loss and left tinnitus persisted. After discussion of the case between the authors, a decision was made to recommend surgery as recurrent vertigo episodes were frequent and felt very debilitating for the young patient. The patient was contacted for a careful explanation of the pathology, and accurate counselling with regard to the operation and the novelty of the approach. The possibility of shifting the operation to an open approach was also explained to the patient in case the endoscopic exclusive transcanal was not adequate for mass removal or for possible bleeding control. The patient eventually agreed to the surgical removal and the operation was performed in March 2012 by the two experienced surgeons (D.M., L.P.).

Surgical Operation

During surgery 0° and 45° rigid endoscopes (Karl Storz, Tuttlingen, Germany) were used, 15 cm in length and 3 mm or 4 mm in diameter. An AIDA three-chip high-resolution monitor and camera (Karl Storz, Tuttlingen, Germany) were used for all of the procedures.

A circumferential tympanomeatal flap was created in the external auditory canal (EAC) with a round knife using the 0° endoscopic view 2 cm from the tympanic membrane; the tympanomeatal flap was then elevated detaching the annulus from the bony ring, and the flap, pedicled on the umbus, was transposed laterally and then detached from the malleus using a microscissors. In this way it was possible to remove all of the eardrum with the meatal skin flap of the EAC, which was preserved in a saline solution (Fig. 2A). To gain optimal surgical access to the entire medial wall of the tympanic cavity, the annulus was drilled to the hypotympanum and the protympanum so that the Eustachian tube was directly visible under 0° endoscopic view. After this procedure good control of the tympanic cavity was achieved, and the ossicular chain, promontory region, protympanum, retrotympanum, and hypotympanum were easily exposed endoscopically (Fig. 2).

Then, using a diamond bur under 0° endoscopic view, the scutum was partially removed, exposing the incudomalleolar joint. The incus was then removed, maintaining the integrity of the stapes. The tensor tendon was cut and the malleus was removed. The stapes was removed and the vestibule was identified with the spherical recess in the saccular fossa (Fig. 2C). The membrane of the round window was clearly identified. Using a Piezosurgery instrument (Mectron, Carasco/Genova, Italy), the lateral aspect of the otic capsule was removed at the cochlear level, identifying the basal, middle, and upper turn of the cochlea. Those labyrinthine spaces were almost completely occupied by the mass (Fig. 2D). A dissection of the intracochlear mass was then performed using the appropriate angled dissectors (Fig. 3A).

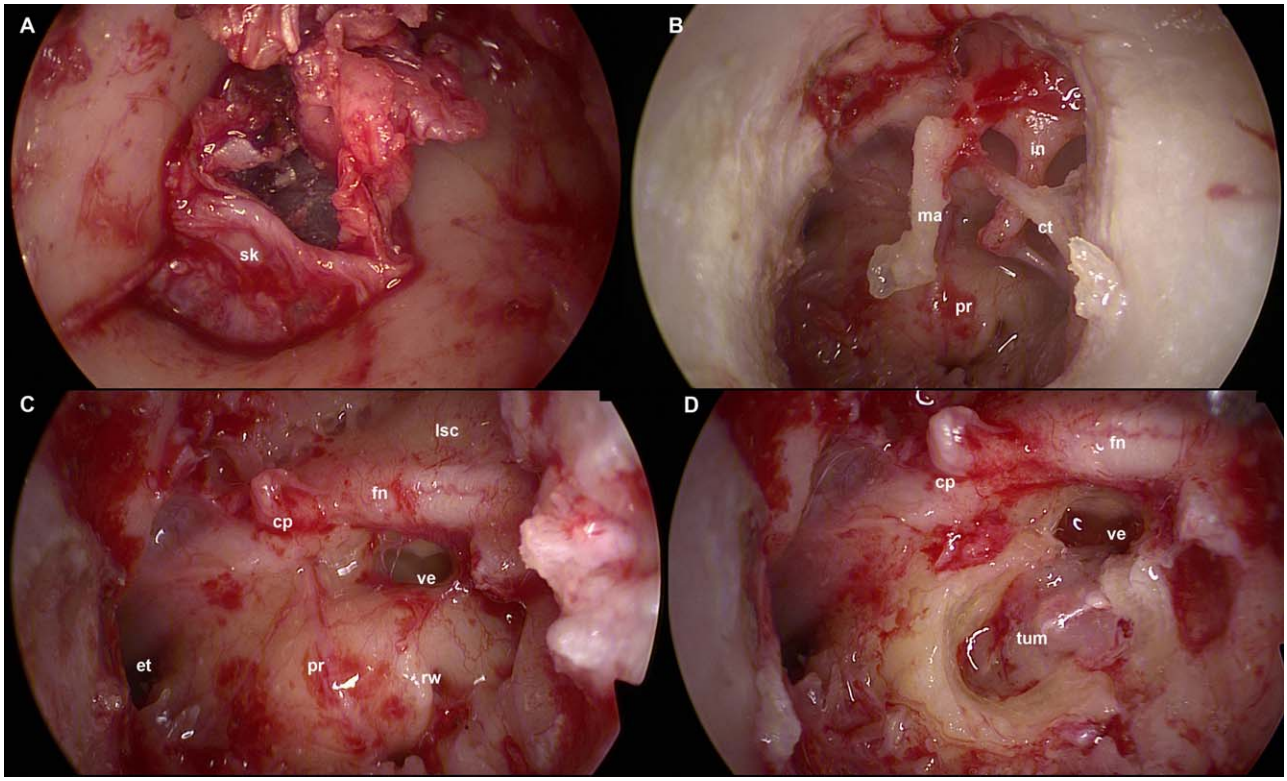

Fig. 2. Surgical stages in the external and middle ear. (A) Degloving of skin and tympanic membrane after a circular incision. (B) Medial aspect of the tympanic cavity is visualized after skin removal. (C) Medial aspect of the tympanic cavity is visualized after skin and ossicular chain removal. Vestibule can be seen through the oval window. (D) Otic capsule opening, with consequent exteriorization of cochlear and vestibular spaces and labyrinthine portion of the neoplasm. cp = cochlear process; ct = chorda tympani; et = Eustachian tube; fn = facial nerve; in = incus; lsc = lateral semicircular canal; ma = malleus; pr = promontory; rw = round window; sk = ear canal skin; tum = tumor; ve = vestibule. [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]

Afterward, using a transcochlear approach, the mass was followed to the IAC, which was opened with exposure of the neuroma into the IAC (Fig. 3B). The dura had a small defect at that level, from which there was a moderate outflow of cerebrospinal fluid (CSF) while removing the pathology. The mass had a close anatomical relationship to the facial nerve, and it was gently dissected, paying careful attention not to damage the nerve. After that an inspection was made of the fundus of the IAC to confirm complete removal of the mass (Fig. 3C). Closure of the IAC was performed with abdominal fat without obliterating the middle ear (Fig. 3D). Cartilage and the perichondrium of the tragus were positioned to isolate the cochlear cavity from the middle ear spaces. Repositioning of the tympanic membrane and external ear canal skin was carried out, reinforcing the graft by positioning a tragal cartilage underlay.

The patient was kept in bed for 3 days after surgery. Immediate postoperative facial function did not show any deficit, but a grade III score (House-Brackmann grading system) was present during the third postoperative day. Postoperative pain was well controlled using intravenous paracetamol. Antibiotic therapy (1 g of cefazoline i.v. twice a day) was administered for 48 hours after surgery. During the first postoperative day, the patient complained of dizziness; and a second-degree nystagmus appeared right-beating. The vertiginous symptoms gradually improved until complete disappearance on the fifth postoperative day. Paralysis of the facial nerve was treated with intravenous corticosteroid therapy for 8 days (20 mg of methylprednisolone twice a day for 4 days, then once a day for a further 4 days). During hospitalization, facial nerve paresis improved to a grade II at discharge.

The patient was discharged on the 9th postoperative day after an overall normal postoperative course. The CT scan at 3-month follow-up showed a normal outcome of the endoscopic transcochlear surgery on the left side (Fig. 4A, B). Postoperative MRI, also after 3 months, did not show pathological enhancement at the site of the surgery, confirming the complete removal of the pathology (Fig. 4C, D).

The facial nerve recovered completely and regained normal function (Fig. 5A, B). The tympanic membrane, visualized by otoendoscopy at the 3-month follow-up, showed a regular outcome for drum reconstruction, without signs of perforation or CSF leakage (Fig. 5C).

DISCUSSION

Retrosigmoid, transmastoid-translabyrinthine, and middle cranial fossa approaches are the most widely

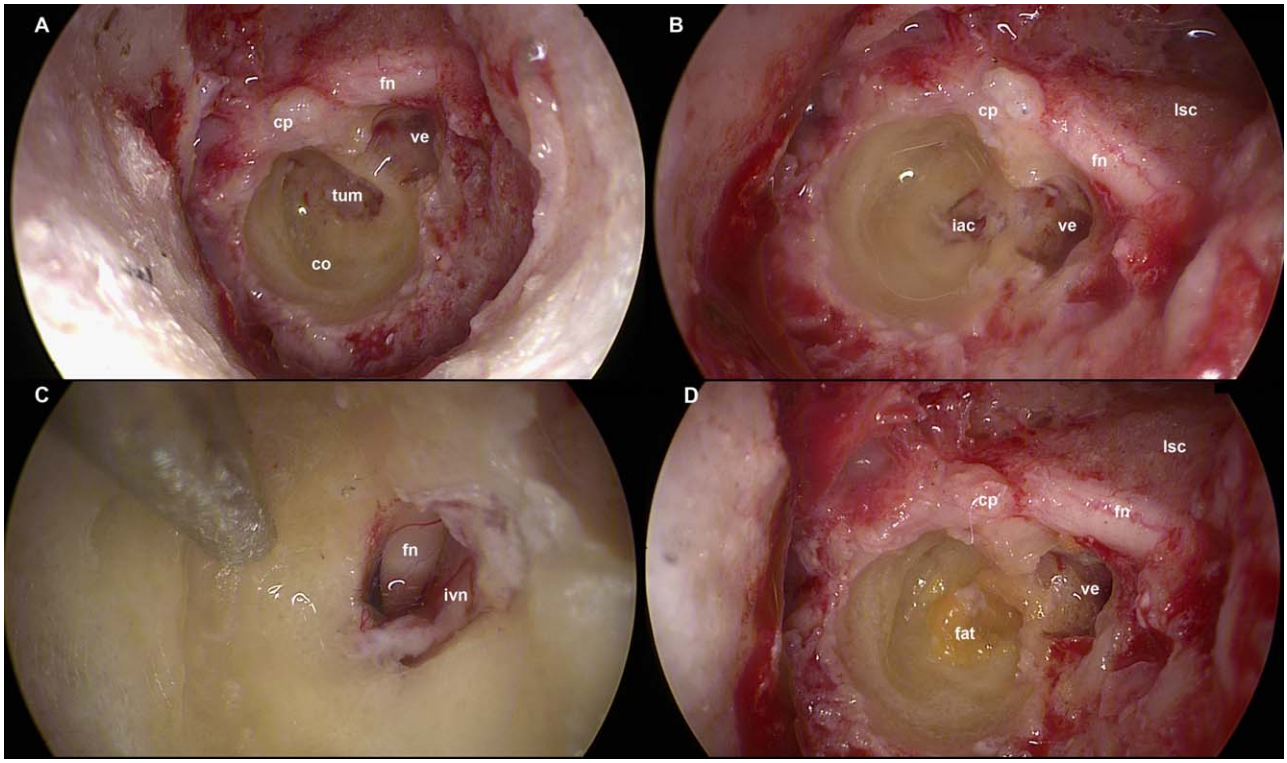

Fig. 3. Surgical stages in the internal ear and internal auditory canal. (A) Appearance of surgical cavity after removal of the cochlear portion of the neuroma. (B) The last piece of tumor, extending into the internal auditory canal, is removed. (C) Closer view of the previous image, showing the fundus of the internal auditory canal and the relationship with the facial nerve at this level. (D) Surgical cavity after positioning of the abdominal fat graft at the level of the fundus of the internal auditory canal. cp = cochlear process; ct = chorda tympani; et = Eustachian tube; fat = fat graft; fn = facial nerve; in = incus; ivn = inferior vestibular nerve; lsc = lateral semicircular canal; ma = malleus; pr = promontory; rw = round window; sk = ear canal skin; tum = tumor; ve = vestibule. [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]

used in the removal of IAC pathology, in particular, in acoustic neuroma (AN) surgery. The choice of the right approach depends on factors such as the surgeon's preferences and habits, dimensions and extent of the pathology, the aim regarding hearing preservation, risk to the facial nerve, and postoperative complications.[1] The middle cranial fossa approach is classically recommended in the case of small Ans: [1] it accesses the IAC superiorly, drilling on the superior aspect of the petrous bone. It guarantees limited access to the cerebellopontine angle (CPA) so that it cannot be used in large acoustic neuromas extending widely outside the IAC. The retrosigmoid approach guarantees optimal control of the CPA, although its control of the canal requires drilling of the posterior aspect of the petrous bone; and in most cases control of the fundus is difficult to obtain.[1] When hearing preservation is attempted, the retrosigmoid and middle cranial fossa approaches are chosen,[9] as occurs frequently for small ANs. Translabyrinthine approaches result in a loss of hearing and extensive drilling of the petrous bone, while they guarantee a good exposure of both IAC and CPA.[1] Although recently some minimally invasive approaches have been recorded, mainly applied to retrosigmoid procedures (i.e., keyhole retrosigmoid approaches),[10] all of these approaches necessitate wide external incisions and soft tissue dissection, craniotomy (retrosigmoid, middle cranial fossa), extensive temporal

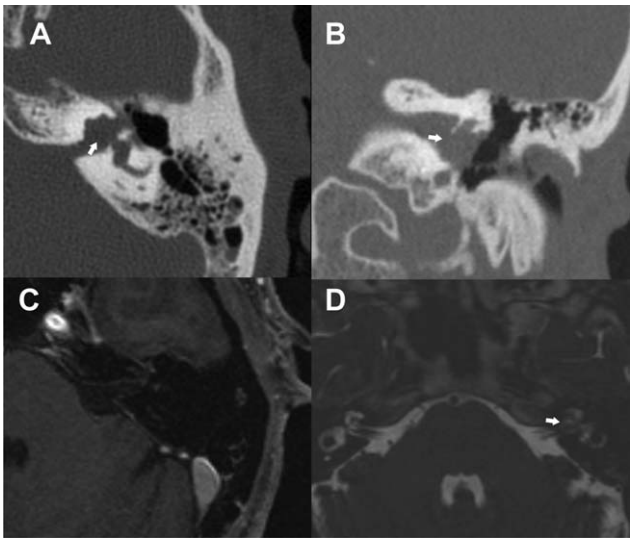

Fig. 4. Postoperative radiologic evaluation. (A, B) At high resolution computed tomography (HRCT) scan (axial and coronal view), the outcomes of surgery are visualized, with apparent communication (arrow) between cochlear spaces and the internal auditory canal. The middle ear cavity is well aerated. (C) Axial T1-weighted MRI with gadolinium. The internal auditory canal can be visualized (arrow) without pathologic enhancements. (D) Axial T2-weighted MRI. The surgical outcomes are visualized, with fat graft in place (arrow), separating the subdural spaces of the internal auditory canal from the cochlea.

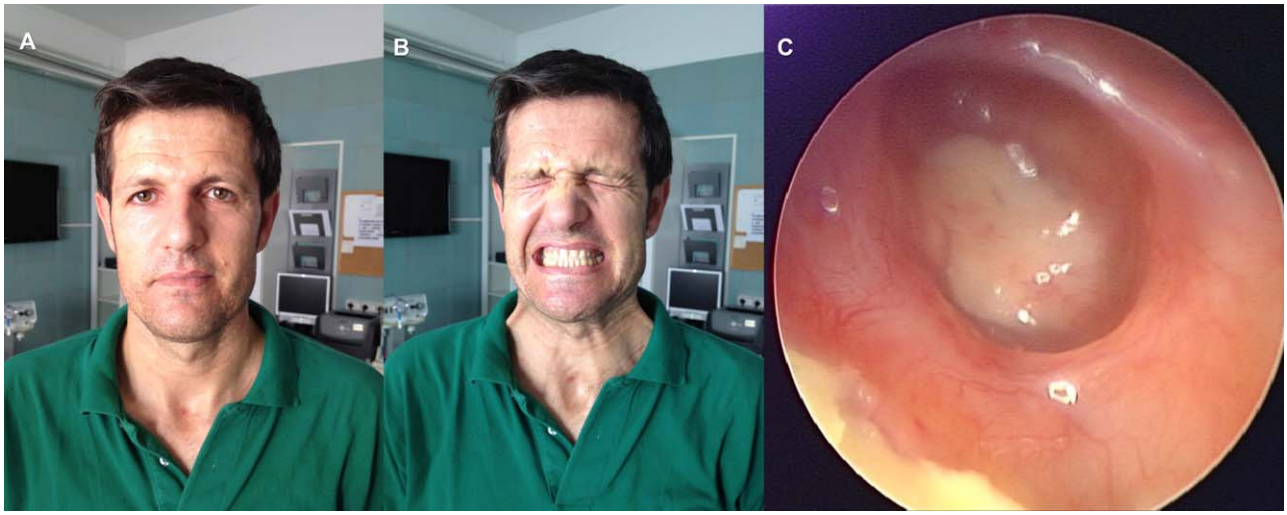

Fig. 5. Facial nerve outcome at 3-month follow-up (A: static; B: dynamic). (C) Otoendoscopic evaluation of tympanic membrane at 3-month follow-up. The cartilage graft is clearly visible through the membrane. [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]

bone drilling and tissue removal (translabyrinthine), cerebellopontine angle exposure by cerebellar retraction (retrosigmoid), or temporal lobe retraction (middle cranial fossa), and cannot really be considered minimally invasive.

As mentioned above, the introduction of middle ear endoscopic surgery is already drastically changing the anatomical and surgical concepts of middle ear surgery. By improving our knowledge and manual skills, and with the evolution of this type of new otologic technique, its application in lateral skull base surgery from the start seemed quite natural to the present authors. For this, by studying the endoscopic anatomy and procedures in cadavers, dissection of the middle ear was gradually completed until the endoscopic anatomy of first the labyrinth and then the IAC were thoroughly known. Some key points of the procedure are herein pointed out.

The first step of the procedures resembles middle ear approaches described in the past for cholesteatoma treatment, or for exploration and study of the middle ear.[5–7,11] In the endoscopic approach to the IAC, the skin of the EAC must be removed en bloc as a glove finger to record the occurrence of the bony EAC during procedures. With removal of the stapes and by closing the optic tip to the oval window, it is possible to visualize a portion of the medial aspect of the vestibule; in particular, in most cases the spherical recess in the saccular fossa can be seen through the oval window. The spherical recess represents an important landmark for further steps since it is the exact place where the inferior vestibular nerve fibres are attached, representing a kind of door for the IAC entrance procedures.

In the authors' experience, the intrameatal facial nerve lies just medially and anteriorly with respect to this opening. After removal of the lateral aspect of the otic capsule by drilling outward from the oval window, it is possible to obtain complete access to the whole medial aspect of the vestibule until the basal turn of the cochlea can be visualized. The medial wall of the vestibule represents the starting point for access to the IAC, just as occurs in the classical translabyrinthine approach. The medial turn of the cochlea certainly represents an important landmark for the intralabyrinthine facial nerve as this segment of the facial nerve runs just superiorly to the cochlea.

Indications for the type of surgery are currently very limited; that is, pathology involving the anterior labyrinth and/or the most lateral portion of the IAC. It is mandatory that these procedures be performed by very experienced professionals, with years of training in endoscopic middle ear procedures. The main problem the authors would like to underline is that in cases of some anterior inferior cerebellar artery (AICA) branches or internal auditory artery bleeding, hemostasis would be very difficult to achieve in inexperienced hands; space is very limited compared to standard procedures. This issue is frequently emphasized by scientific community discussing risks of exclusive endoscopic retrosigmoid procedures (by "key-hole" technique). At present we also consider it to be the main issue and possible criticism to our approach. It must be underlined that a careful preoperative magnetic resonance (MR) study or CPA vascularization is mandatory in that kind of surgery; and in cases that unfavorable vascular anatomy was noticed, the approach should not be recommended. The debate about natural orifice transluminal surgery (NOTES) is nowadays in every sector of surgery.[12] We should also consider this kind of approach not only a minimally invasive procedure, but also a NOTES procedure. Although the approach described should not be considered a standard technique, but rather an innovative approach, the likely evolution of instruments or the introduction of existing technologies to this surgery (i.e., robotic surgery) could enormously expand the indications, possibly completely changing the traditional approaches and procedures to lateral skull base surgery.

CONCLUSION

The transcanal exclusive endoscopic approach proved to be successful for CS removal involving the fundus, IAC, and cochlea. Potential future application of this kind of approach in lateral skull base surgery will depend on the development of technology and surgical and anatomical refinements.

BIBLIOGRAPHY

1. Bennett M, Haynes DS. Surgical approaches and complications in the removal of vestibular schwannomas. *Otolaryngol Clin North Am* 2007;40:589–609.
2. Magnan J, Chays A, Lepetre C, Pencroffi E, Locatelli P. Surgical perspectives of endoscopy of the cerebellopontine angle. *Am J Otol* 1994;15:366–370.
3. Thomassin JM, Korchia D, Doris JM. Endoscopic guided otosurgery in the prevention of residual cholesteatomas. *Laryngoscope* 1993;103:939–943.
4. Presutti L, Marchioni D, Mattioli F, Villari D, Alicandri-Ciufelli M. Endoscopic management of acquired cholesteatoma: our experience. *J Otolaryngol Head Neck Surg* 2008;37:481–487.
5. Marchioni D, Alicandri-Ciufelli M, Molteni G, Genovese E, Presutti L. Endoscopic tympanoplasty in patients with attic retraction pockets. *Laryngoscope* 2010;120:1847–1855. doi: 10.1002/lary.21069.
6. Marchioni D, Alicandri-Ciufelli M, Piccinini A, et al. Surgical anatomy of transcanal endoscopic approach to the tympanic facial nerve. *Laryngoscope* 2011;121:1565–1573.
7. Marchioni D, Villari D, Alicandri-Ciufelli M, Piccinini A, Presutti L. Endoscopic open technique in patients with middle ear cholesteatoma. *Eur Arch Otorhinolaryngol* 2011;268:1557–1563.
8. Tarabichi M. Endoscopic management of limited attic cholesteatoma. *Laryngoscope* 2004;114:1157–1162.
9. Staecker H, Nadol JB Jr, Ojeman R, Ronner S, McKenna MJ. Hearing preservation in acoustic neuroma surgery: middle fossa versus retrosigmoid approach. *Am J Otol* 2000;21:399–404.
10. Shahinian HK, Ra Y. 527 fully endoscopic resections of vestibular schwannomas. *Minim Invasive Neurosurg* 2011;54:61–67. Epub 2011.
11. Marchioni D, Alicandri-Ciufelli M, Piccinini A, Genovese E, Presutti L. Inferior retrotympanum revisited: an endoscopic anatomic study. *Laryngoscope* 2010;120:1880–1886. doi: 10.1002/lary.20995.
12. Berney CR. Natural orifice transluminal endoscopic surgery: Where should we draw the line? *Ann Surg* 2011;254:1081–1083. doi: 10.1097/SLA.0b013e31823ad063.