*Original Research*

# Endoscopic-Assisted Cochlear Implantation in Children with Malformed Ears

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## Abstract

*Objective.* Complex middle and inner ear malformations are considered an important limitation for cochlear implant (CI) with traditional microscopic techniques. The aim of the present study is to describe the results of the endoscopic-assisted CI procedure in children with malformed ears.

*Study Design.* Case series with chart review of consecutive patients.

*Setting.* Two tertiary referral centers: University Hospital of Verona and University Hospital of Modena, Italy.

*Subjects and Methods.* In total, 25 children underwent endoscopic-assisted cochlear implantation between January 2013 and January 2018. The audiologic and neuroradiologic assessment showed profound hearing loss and malformation of the middle and inner ear in all children. A complete review of anatomic features, surgical results, and audiologic outcomes was performed. The surgical technique is described step-by-step, and the outcomes are detailed.

*Results.* All patients (mean age, 3.6 years; range, 2.8-9 years) underwent a transattical/endoscopic-assisted CI procedure. All children showed varying degrees of auditory benefit, as measured by routine audiometry, speech perception tests, and Categories of Auditory Performance scores (mean, 6). No immediate or late postoperative complications were noted.

*Conclusion.* The endoscopic-assisted approach proved to be successful in cochlear implantation. The direct visualization and magnification allow (1) exploration of the tympanic cavity; (2) confirmation of all anatomic features, with strict control of the course of the facial nerve, round window area, and inner ear; and (3) performance of the cochleostomy with adequate insertion of the array.

## Keywords

endoscopic ear surgery, ear malformations, round window, cochlear implantation, children

Complete visualization of the round window (RW) during cochlear implantation is mandatory for correct placement of the array in the cochlea and to minimize intracochlear damage. In cases of middle and inner ear malformations, access to the promontory and RW via standard posterior tympanotomy can be limited. Under these conditions, not only does insertion of the electrodes into the cochlea with a microscope involve risk of damage to the facial nerve (FN), but it is also impossible to approach the cochlea due to a partially or totally hidden RW niche and membrane.[1]

It was recently demonstrated that the endoscopic approach enables excellent visualization and exploration of the RW niche during ear surgery, especially in patients with middle and inner ear malformations.[2-5]

This study represents the largest case series of patients with ear malformations who have undergone endoscopic-assisted cochlear implantation. The aim of this study is to explore the utility of the endoscope in visualization of the RW membrane (RWM) during cochlear implant (CI) and to check the adequate insertion of the electrodes into the scala tympani (ST) in patients with middle and inner ear malformations.

## Materials and Methods

### Subjects

We carried out a chart review of consecutive patients who underwent endoscopic-assisted CI surgery at 2 tertiary referral centers (University Hospital of Verona, University Hospital of Modena, Italy) between January 2013 and

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January 2018. The mean follow-up was 24.6 months (range, 6-58 months). Endoscopic assistance was planned for all patients scheduled for CI and with certain or suspected middle and inner ear malformations.

The study population consisted of 25 pediatric patients (<16 years of age) affected by profound hearing loss and malformation of the middle and inner ear (**Table 1**). Patients with no ear malformations, as reported at the preoperative neuroradiologic evaluation, were not included in this study.

The presence of anatomic malformations was detected with preoperative temporal bone high-resolution computed tomography (**Figure 1**) and magnetic resonance imaging.[6] The complexity of inner ear malformations was classified according to recent criteria reported in the literature. The classification by Sennaroğlu and Bajin[7] is based on neuroradiologic findings and describes 6 types of inner ear malformation: complete labyrinthine aplasia, rudimentary otocyst, cochlear aplasia, common cavity, cochlear hypoplasia, and incomplete partition of the cochlea.

The accessibility of the RWM was classified according to the St Thomas' Hospital classification.[1] This illustrates 4 types of RWM; type I, a fully exposed RWM; type II, partial exposure; type IIa, exposure >50% but <100%); and IIb, exposure <50% but >0%. In type III, the RWM cannot be identified at all, even after best surgical effort.

Before surgery, comprehensive audiologic examinations with routine or behavioral audiometry, speech perception tests, and Categories of Auditory Performance (CAP) score were performed for all patients. The CAP score comprises a hierarchical scale of auditory perceptive ability ranging from 0 (displays no awareness of environmental sounds) to 7 (can use the telephone with a familiar talker).[8]

All children showed profound hearing loss with no verbal production.

## Surgery

The endoscopic-assisted CI procedures were performed by 2 surgeons (L.P. and D.M.). All procedures were recorded and analyzed postoperatively. Intraoperative anatomic findings were reported, as well as any surgical complications encountered during the procedure. Classification of ear malformations and accessibility of the RWM was conducted by 1 author (M.C.) according to the classifications described earlier.

The endoscopic-assisted CI procedure was performed in 2 steps: (1) endoscopic step with a 0° endoscope, 3 mm in diameter, 14 cm in length, via a transcanal route; (2) microscopic step via the transmastoid transattical route.

The use of the transattical route was chosen for these patients to minimize the risk of FN injuries during surgery, which is higher in the case of complex ear malformations.

The details of these surgical steps are reported elsewhere[9] and are briefly summarized here.

*Endoscopic Step.* The endoscope was inserted through the external auditory canal, and a tympanomeatal flap was lifted anteriorly to enter the tympanic cavity. The course of the FN, the ossicular structures, the RW niche, and the promontory was visualized. The RW niche was identified and its accessibility evaluated to visualize the ST clearly (**Figure 2**). After the tegmen was drilled with a diamond burr, the RWM was exposed and carefully opened with a microhook, creating access to the ST. A piece of Gelfoam was then placed on the cochleostomy site. When the RW was not present, a promontorial endoscopic cochleostomy was performed with careful opening of the ST, keeping the FN course in direct sight.

*Microscopic Step.* A retroauricular skin incision was performed; after identification of the plane of the temporal muscle fascia, a posterior periosteal flap was then created and elevated uncovering the mastoid bone. The skin of the posterior wall of the external auditory canal was elevated to detect the tympanic cavity. A mastoidectomy was performed, and the antrum, the atticus, and the incudomalleolar joint were exposed. A wide connection between the posterior epitympanum and the mesotympanic spaces was created by removal of the incus. The receiver-stimulator of the implant was fixed and covered under the temporalis muscle.

The array pushed through the corridor from the epitympanum into the mesotympanum was gently inserted into the cochleostomy. A small piece of temporalis fascia was used with fibrin glue to seal the cochleostomy. The tympanomeatal flap was replaced and the external auditory canal packed with fragments of Gelfoam. The subperiosteal flap covering the CI receiver body and the retroauricular skin incision was sutured in layers.

### Ethical Considerations

This study was approved by the Research Ethics Committee of Verona University Hospital and the Research Ethics Committee of Modena University Hospital. Informed consent for surgery was obtained from all of the children's parents.

## Results

Between January 2013 and January 2018, 25 consecutive patients (13 female and 12 male; mean age, 3.6 years; range, 2.8-9 years) underwent CI surgery with an endoscopic-assisted procedure at 2 tertiary referral hospitals (University Hospital of Verona, University Hospital of Modena).

The main middle and inner ear malformations observed are detailed in **Table 1**. In all children, the endoscopic-assisted approach allowed the anatomic conformation of the tympanic cavity to be recognized, as well as the morphology of the ossicular chain and the position of the FN, but above all, it permitted clear identification of the RWM and the ST after an RW cochleostomy.

Twenty-two children underwent unilateral cochlear implantation, and 3 had a simultaneous bilateral CI. In 1 child, a simultaneous contralateral auditory brainstem implant was performed with a translabyrinthine approach.

An anomalous course of the FN was found in 24 of the 25 children (27 ears), and malformations of the ossicular chain were reported in 20 patients (23 ears). Exposure of the RWM was limited in all children: in 23 children (25

**Table I.** Summary of Patients' Features.

| No. | Age, y | Disease | Ossicular Chain | FN Course | Cochlea/Vestibule[a] | Round Window[b] | IAC | CI |
|---|---|---|---|---|---|---|---|---|
| 1 | 4 | CHARGE syndrome | Malformed | Abnormal | IP II | Abnormal, covered by FN (IIb) | Hypoplastic | MED-EL |
| 2 | 3 | Nonsyndromic | Normal | Abnormal | Cochlear hypoplasia | Abnormal site (IIb) | Normal | Cochlear |
| 3 | 9 | Bilateral microtia | Malformed | Abnormal | IP II | Abnormal site (IIb) | Normal | MED-EL |
| 4 | 3 | Nonsyndromic | Malformed | Abnormal | Cochlear hypoplasia | Abnormal prolapse tegmen (IIb) | Normal | Cochlear |
| 5 | 5 | Nonsyndromic | Normal | Normal | IP I | Normal (IIb) | Normal | Cochlear |
| 6 | 3 | CHARGE syndrome | Malformed | Abnormal | Cochlear hypoplasia | Round window covered by bone (III) | Hypoplastic | Cochlear |
| 7 | 5 | Nonsyndromic | Normal | Abnormal | IP II | Abnormal site (IIb) | Normal | Cochlear |
| 8 | 3 | Bilateral atresia auris | Malformed | Dehiscent | IP I | Abnormal site (IIb) | Hypoplastic | Cochlear |
| 9 | 3 | Nonsyndromic | Malformed | Abnormal | IP I | Covered by FN (IIb) | Normal | Cochlear |
| 10 | 3 | Bilateral atresia auris | Malformed | Abnormal | IP II | Abnormal site (IIb) | Hypoplastic | MED-EL |
| 11 | 5 | Nonsyndromic | Malformed | Abnormal | Cochlear hypoplasia | Abnormal site (IIb) | Hypoplastic | MED-EL |
| 12 | 4 | CHARGE syndrome | Malformed | Abnormal | Cochlear hypoplasia | Partially covered by bone (IIb) | Hypoplastic | MED-EL |
| 13 | 5 | Nonsyndromic | Normal | Abnormal | IP II | Abnormal site (IIb) | Normal | Cochlear |
| 14 | 3 | Bilateral atresia auris | Bilaterally malformed | Bilateral dehiscent | IP I right, IP II left | Bilateral abnormal site (IIb) | Hypoplastic right and left | Cochlear right and left |
| 15 | 2 | Nonsyndromic | Malformed | Abnormal | IP I | Abnormal site (IIb) | Normal | Cochlear |
| 16 | 3 | Bilateral atresia auris | Bilaterally malformed | Bilateral abnormal | IP II bilateral | Bilateral abnormal site (right IIb, left III) | Hypoplastic right and left | Cochlear right and left |
| 17 | 2 | Nonsyndromic | Normal | Abnormal | Cochlear hypoplasia | Abnormal site (IIb) | Hypoplastic | MED-EL |
| 18 | 3 | CHARGE syndrome (CI reintervention) | Malformed, cholesteatoma | Abnormal | Cochlear hypoplasia | Abnormal site (IIb); array enveloped by cholesteatoma | Hypoplastic | Cochlear |
| 19 | 8 | CHARGE syndrome | Malformed | Abnormal | Cochlear hypoplasia | Abnormal site (IIb) | Hypoplastic | MED-EL |
| 20 | 2 | Syndromic (cardiac situs inversus, septum pellucidum vergae cavum) | Bilaterally malformed | Bilateral abnormal | IP II bilateral | Abnormal site (IIb) | Normal right, hypoplastic left | MED-EL, MED-EL (ABI) |
| 21 | 3 | CHARGE syndrome | Bilaterally malformed | Bilateral abnormal | IP III right, IP II left | Bilateral abnormal site (IIb) | Hypoplastic right and left | MED-EL right and left |
| 22 | 2 | Nonsyndromic | Malformed | Abnormal | Cochlear hypoplasia | Abnormal site (IIb) | Hypoplastic | MED-EL |
| 23 | 2 | Nonsyndromic | Malformed | Abnormal | IP III | Abnormal site (IIb) | Hypoplastic | MED-EL |
| 24 | 3 | CHARGE syndrome | Malformed | Abnormal | Cochlear hypoplasia | Abnormal site (IIb) | Hypoplastic | Cochlear |
| 25 | 3 | Bilateral microtia | Malformed | Abnormal | IP I | Abnormal site (IIb) | Normal | Advanced Bionics |

Abbreviations: ABI, auditory brainstem implant; CI, cochlear implant; FN, facial nerve; IAC, internal auditory canal; IP I, incomplete partition type I; IP II, incomplete partition type II; IP III, incomplete partition type III.
[a]Sennaroğlu classification.[7]
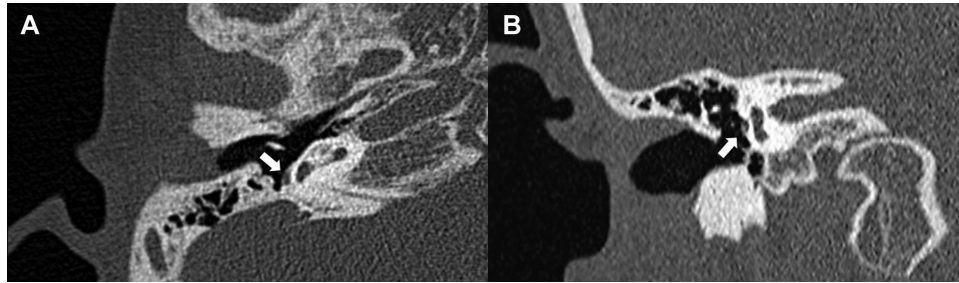[b]St Thomas' Hospital classification.[1]

**Figure 1.** Patient 1. Preoperative computed tomography scan, bone window: (A) axial and (B) coronal views of the right ear. Anomalous course of facial nerve (white arrow) runs over the promontory and covers the round window.
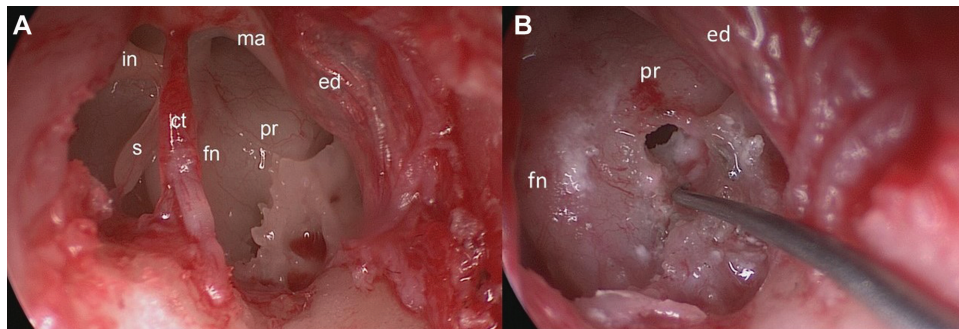


**Figure 2.** Patient 1. Endoscopic view, right ear: (A) examination of the malformed middle and inner ear and (B) promontorial cochleostomy. ct, chorda tympani; ed, eardrum; fn, facial nerve; in, incus; ma, malleus; pr, promontory; s, stapes.

ears), less than half of the RWM was visible after drilling the niche (St Thomas' Hospital classification type IIb), and in 2 children (2 ears), the RWM was not visible (type III).

The preoperative radiologic evaluation predicted an inadequate surgical RW exposure in only 7 of 25 children (8 ears, 28%), in addition to a significant degree of unfavorable cochlear position; in 18 of 25 children (20 ears, 72%), the accessibility of the RW could not be predicted with certainty before surgery. In 24 of 25 children (27 ears), an RWM cochleostomy was performed, and 1 child with absent RW required a promontorial endoscopic cochleostomy.

Thirteen children (15 ears, 2 patients with bilateral simultaneous surgery) underwent implantation with a Cochlear device (Cochlear, Macquarie University, New South Wales, Australia); 11 children (12 ears, 1 child with bilateral simultaneous CI and another with a contralateral simultaneous auditory brainstem implant) received a MED-EL implant (Innsbruck, Austria); and 1 patient underwent rehabilitation with a HiRes 90K Advantage device (Advanced Bionics, Valencia, California).

### Complications

All patients were discharged from hospital on the day after surgery. No intra- or postoperative complications were observed in this series. In particular, no intraoperative gusher or FN damage was found in any child.

### Audiologic Results

The mean CAP score[8] was 6 (range, 5-7) at last follow-up.

## Discussion

Cochlear implantation outcomes vary widely among patients. This variability is due to several factors, such as differences in patient age at implantation,[10] duration of deafness,[11] coding strategies,[12] and, more recently, surgical technique, in relation to normal or distorted anatomic condition of the ear.[4,13] Patients with electrodes in a proper placement along the ST, as a consequence of correct vector insertion coaxial with the centerline of the ST,[14] enjoy better hearing outcomes.[15] If this direction is not maintained, the implant not only gives lower auditory performance but may also penetrate the basilar membrane directly or after glancing off the wall of the cochlea farther along the course of the basal turn.[14]

The surgeon may attempt to ensure a coaxial insertion by estimating the course of the ST of the basal turn, observing its lumen through the cochleostomy, but this view is necessarily limited or not possible with a microscope, particularly in children with middle and inner ear malformations.[5,14] The endoscopic-assisted technique allows this obstacle to be overcome. The advantages of this technique in middle ear surgery, particularly in the evaluation of the RW region, are now well known[16] and are not discussed in this article. This study aimed to verify the main benefit of endoscopic-assisted CI in improving the visibility of the RW region for correct insertion of the electrodes into the ST of children with malformed ears. In the literature, the percentage of totally hidden or absent RW in CI surgery varies from 4% to 14% in children with malformed ears[1,5,17,18]; similar data

(8%) were found in our case series. The surgical exposure of the RW cannot always be reliably predicted with the preoperative temporal bone computed tomography scan; the use of the endoscope in these conditions is essential to visualize the RW and to perform a correct cochleostomy.[5] In our experience, the difficult surgical exposure of the RWM was evident in all children, and in 18 subjects (19 ears), it could not be predicted preoperatively. These findings are similar to those reported in the literature.[5]

The endoscopic technique easily permitted the morphology of the RWM to be determined and the fustis, the area concamerata, and the subcochlear canaliculi to be visualized in all conditions[19]; it also allowed the orientation of the ST of the basal turn to be clearly identified, providing useful information on the spatial orientation of the modiolus and ensuring a correct placement and advancement of the electrodes in the cochlea.

However, the direct line of sight obtained by the microscope through the posterior tympanotomy may not provide sufficient surgical exposure, and sometimes, it may be impossible to identify the RWM. The visualization and accessibility of the RWM evaluated under microscopic and endoscopic views demonstrated a significant improvement with the endoscope.[17] The possibility offered by the endoscope to visualize the RW and evaluate its orientation is important in children and in normal auditory anatomy, due to the more obtuse angle between the cochlea and the temporal bone in the pediatric population.[20,21]

Intra- or postoperative complications were not observed in any of our patients, and we were able to obtain a proper and adequate insertion of the array in the cochlea through an RW cochleostomy in 27 of 28 ears and through a promontorial cochleostomy in 1 ear. In none of the cases did we need to change the surgical approach during the surgical session or to abandon the surgery, as sometimes reported in the literature.[22,23]

The drawbacks of the endoscopic-assisted procedure consist mainly of the learning curve required for the surgeon to gain confidence with this one-handed technique; moreover, the transcanal approach increases the risk of injury to the tympanomeatal flap, including tympanic membrane perforation with subsequent risk of infection through the defect. Furthermore, management of intraoperative major complications cannot be achieved with an endoscopic exclusive technique; for example, in case of intraoperative gusher, the sealing of the RW is required, and in some cases, a subtotal petrosectomy must be performed.

The transattical approach for cochlear implantation is an efficient alternative to the standard posterior tympanotomy procedure. It showed similar results in terms of complication rates and CI performance. The electrode's integrity is not significantly affected by its different position and angles of insertion when compared with the standard approach.[24] Nevertheless, the removal of the incus during the transattical approach limited the use of the stapedial reflex for estimation of C-level (ie, comfort or maximal stimulation level); this is a significant disadvantage in young children,

especially those with developmental delay associated with their syndrome, wherein their ability to express their perceptions is limited.

The good outcomes obtained in our patients in terms of auditory rehabilitation (mean CAP score, 6) are promising and confirm the utility of the endoscope for correct insertion of the electrodes into the ST of children with malformed middle and inner ear.

The first advantage offered by the endoscopic-assisted technique is magnification—specifically, the enlargement of the visual field as the endoscope approaches the object of interest. The second is the unique ability of angled endoscopes to visualize "around the corner." Both features contribute to obtaining a panoramic view of the anatomic structures, which may not be possible with the direct line of vision of the microscope; this possibility is particularly important for accessibility to the RW area and for proper insertion of the electrodes into the ST.[25,26] Since some electrodes are particularly flexible and elastic whereas others may require removal of the stylet with one hand and insertion with the other, it is an unnecessary challenge to try to insert the electrode while one hand is holding the endoscope. Therefore, we do not recommend routinely inserting the electrode under endoscopic view. The transcanal corridor is preferred since, as is well known, it offers the best frontal direct view of the RW area.[2,17] However, the endoscopic transcanal approach for cochlear implantation[2] showed several disadvantages: higher risk of extrusion of electrodes, chronic infections, and acquired cholesteatoma. In addition, a transcanal approach would provide entry to the basal turn of the cochlea at a significant angle to the axis of the segment of the basal turn.[27]

The combined technique described here is recommended as an auxiliary approach to overcome the abnormal anatomic conditions of a malformed middle and inner ear in children and to perform an effective cochleostomy.

## Conclusions

Based on our personal experience illustrated here, the use of a transattical/endoscopic-assisted approach could be helpful as an auxiliary technique for cochlear implantation in children with anatomic malformation of the middle and inner ear. As compared with traditional microscopic approaches, it allows magnification of middle ear structures with the main advantage of direct view of the RWM and FN, even when complex malformations limit visualization with the microscope. Of course, further studies will be necessary, and long-term validation and lengthy follow-up are required; however, the surgical and functional results obtained in this series of children are promising.

### Author Contributions

**Marco Carner**, conception of the work, acquisition, analysis, and interpretation of data for the work, revision of work for important intellectual content, final approval of version to be published, agreement to be accountable for all aspects of work; **Andrea Sacchetto**,

acquisition, analysis, and interpretation of data for the work, revision of work for important intellectual content, final approval of the version to be published, agreement to be accountable for all aspects of work; **Luca Bianconi**, acquisition, analysis, and interpretation of data for the work, revision of work for important intellectual content, final approval, agreement to be accountable for all aspects of work; **Davide Soloperto**, analysis and interpretation of data for the work, revision of work for important intellectual content, final approval of version to be published, agreement to be accountable for all aspects of work; **Luca Sacchetto**, analysis and interpretation of data, revision of work for important intellectual content, final approval, agreement to be accountable for all aspects of work; **Livio Presutti**, conception of the work, revision of work for important intellectual content, final approval, agreement to be accountable for all aspects of work, **Daniele Marchioni**, conception and design of the work, revision of work for important intellectual content, final approval, agreement to be accountable for all aspects of work.

## Disclosures

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