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Endoscopic-assisted cochlear implant procedure in CHARGE syndrome: Preliminary report

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ABSTRACT

The endoscopic approach demonstrated to be useful for cochlear implantations of children with CHARGE syndrome. It allows the surgeon to perform a direct 'safe' cochleostomy in difficult anatomical conditions. Three children with CHARGE syndrome underwent endoscopic-assisted cochlear implant surgery at the Tertiary University Referral Center of Modena and Verona between January 2014 and September 2015. A review of clinical data and videos from the operations was made. All procedures were re-analyzed and codified. Three children, all females (mean age 4.6 years; range: 3–7 years) underwent surgery. Two primary surgical procedures and one revision surgery, for secondary cholesteatoma, were performed. CT scans demonstrated complex malformations of middle and inner ear with anomalous course of the facial nerve. In all subjects, a transcanal endoscopic cochleostomy was performed; no immediate or late postoperative complications were observed. Discharge from hospital was the day post-surgery. The current mean follow-up is 12.5 months (range: 8–19 months).

Introduction

Ear abnormalities and hearing loss are common in children with CHARGE syndrome [1]. Both conductive hearing loss, due to chronic otitis media, cholesteatoma or ossicular chain abnormalities, and sensorineural hearing loss, due to inner ear abnormalities, can be found. CHARGE syndrome is also associated with cranial nerve abnormalities, particularly involving the olfactory, facial, vestibular and cochlear nerves [2]. A small number of these children have profound hearing loss and are eligible for cochlear implant (CI). However, children with CHARGE syndrome have worse audiological outcomes after surgery for several reasons, especially related to the difficult insertion of the array in the malformed site and to the grade of cochlear malformation. In these cases, the correct exposure of the anatomical landmarks is crucial to perform a safe and successful surgical intervention avoiding facial nerve (FN) injury and yielding the correct location of the cochleostomy for array insertion. Furthermore, the high risk of gusher in these patients [3] and the condition of the cochlear nerve in the internal auditory canal (IAC) must be considered before surgery. Recently, an endoscopic-assisted technique has been proposed to overcome the surgical problems of the CI procedure in malformed ear [4]. The aim of this study is to analyze the contribution of the endoscopic approach in CI surgery for the management of children with CHARGE syndrome. Functional results, preoperative (radiological and electrophysiological) assessment and intraoperative findings were also reported.

Materials and methods

Subjects

A chart review of clinical data and videos from the operations of three children affected by CHARGE syndrome which underwent endoscopic-assisted CI surgery was conducted. The surgical procedures were performed at the Tertiary University Referral Center of Modena and Verona between January 2014 and September 2015. A complete audiological and neuro-radiological preoperative assessment was performed to confirm the indications for surgery and to study middle and inner ear status. The informed consent to the surgery was obtained from children's parents. Intraoperative anatomical findings and degree of implant insertion were recorded, as well as any surgical complications encountered during this procedure.

On the basis of the CT and MRI scan findings, all inner and middle ear malformations were analyzed and classified [5]. Patient characteristics are shown in Table 1. Three females (mean age 4.6 years; range: 3–7 years) underwent this endoscopic-assisted surgical procedure, 2 out of 3 required primary surgical procedures (#1; #3); 1 out of 3 patients underwent previous CI in another institution. She was affected by secondary cholesteatoma which enveloped the array and required revision surgery (#2).

In all subjects, an anomalous course of the FN and malformations of inner and middle ear were present, as showed by CT scan (Figure 1, Panels A–D). According to the Sennaroglu and Saatci's classification [5], an incomplete partition type II was revealed in one subject and all presented anomalous conformation of the semicircular canals and a hypoplastic cochlear nerve.

Data were collected regarding: patient demographics and past medical history; type and severity of hearing loss; age at cochlear implantation; language production before surgery; neuroradiological findings; preoperative and intraoperative electrophysiological measurements; complications and language outcomes, including Categories of Auditory Performance scores [6] (CAP). Patient characteristics are shown in Table 1.

Surgical technique

The endoscopic-assisted CI procedure was performed in two steps. The first step was performed trancanally under endoscopic view. The endoscopic magnification allowed to recognize the anatomical conformation of the tympanic cavity and to perform the cochleostomy. The second step was performed under the microscopic view through the trans-mastoid trans-attic route. The array was inserted, through the attic to the mesotimpanum, into the cochleostomy previously created. The details of these steps are as following:

Endoscopic steps

A 0° endoscope, 3 mm of diameter, 15 cm in length, was inserted into the external auditory canal and used to create a tympanomeatal flap that was elevated under endoscopic view, entering into the tympanic cavity (Figure 2, Panel A). The flap was then pulled anteriorly, until the posterior border of the malleus was identifiable. An endoscopic check of the anatomical structures in the tympanic cavity was made. The relationship between the abnormal course of the FN, the ossicular structures, the round window (RW) niche and the promontory was observed endoscopically (Figure 2, Panel B). The RW niche was identified, valuating its accessibility. If the RW niche was not accessible, a diamond burr was used under endoscopic view to drill the tegmen, having a good exposure of the RW membrane to perform an endoscopic cochleostomy. A micro-hook was then used to open the membrane, creating the access to the scala tympani. A piece of gelfoam was placed on the cochleostomy site. When the RW was inaccessible, due to an anomalous course of the FN over the RW niche, hiding it, an endoscopic promontorial cochleostomy was performed. In these cases, the promontorial cochleostomy was useful in order to maintain the FN and RW niche under endoscopic control, avoiding surgical injury (Figure 2, Panel C). The cochleostomy was performed just anteriorly and inferiorly to the RW, carefully opening the scala tympani. A piece of gelfoam was placed on the cochleostomy site (Figure 2, Panel D).

Microscopic steps

A retroauricular skin incision was made, identifying the plane of the temporal muscle fascia; a posterior

Table 1. Data of children with CHARGE syndrome.

Patient (surgery) Side	Age	Ossicular chain	Facial nerve	Cochlea/vestibule	RW	IAC	CI	Intraop. findings	Auditory and verbal outcomes
C.F. Right	4 years	Malformed	Abnormal course	Hypoplastic; absent SC; incomplete partition type II	Abnormal, covered by FN	Normal; hypoplastic CN	Medel	ART Poor waveforms	No expressive sign or verbal language since CI. CAP 6 at the last fu (15 months)
Z.G. Right (revision surgery)	3 years	Malformed (cholesteatoma)	Abnormal course	Hypoplastic; absent SC	Abnormal, covered by FN	Normal; hypoplastic CN	Cochlear	NRT Clear waveforms	No expressive sign or verbal language since CI. CAP 6 at the last fu (5 months)
M.G. Left (second side)	7 years	Malformed	Abnormal course	Hypoplastic; absent SC	Abnormal, covered by FN	Normal; hypoplastic CN	Medel	ART Slight waveforms	No expressive sign or verbal language since CI. CAP 6 at the last fu (5 years with mono CI; 6 mo with bilateral CI)

RW: round window; IAC: internal auditory canal; CI: cochlear implant; SC: semicircular canals; FN: facial nerve; CN: cochlear nerve; ART: auditory nerve response telemetry; NRT: neural response telemetry; CAP: categories of auditory performance.
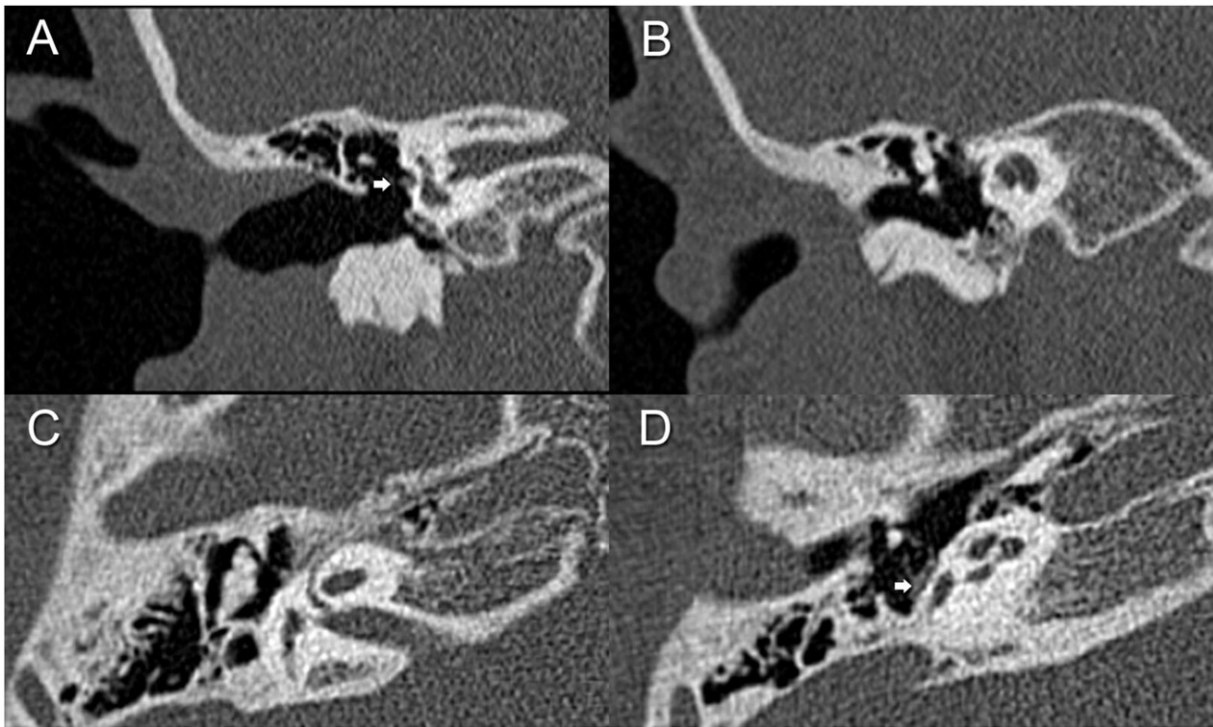
Figure 1. Preoperative CT scan (Patient #3). Panel A. Coronal view: the facial nerve runs over the promontory in anomalous position (white arrow), the stapes is not visible. Panel B. Coronal view: the turns of the cochlea are present; the head of the malleus is fused with the tegmen. Panel C. Axial view: the internal auditory canal has a normal conformation; a hypoplastic canal of cochlear nerve is seen; the labyrinthine portion of the facial nerve runs just posteriorly with respect to the cochlea and in an inferior position with respect to the apical turn of the cochlea, the incudo-malleolar joint looks normal, a hypoplasic labyrinthine is seen. Panel D. Axial view: the facial nerve (white arrow) runs over the promontory, an anomalous incudo-stapedial joint is seen.
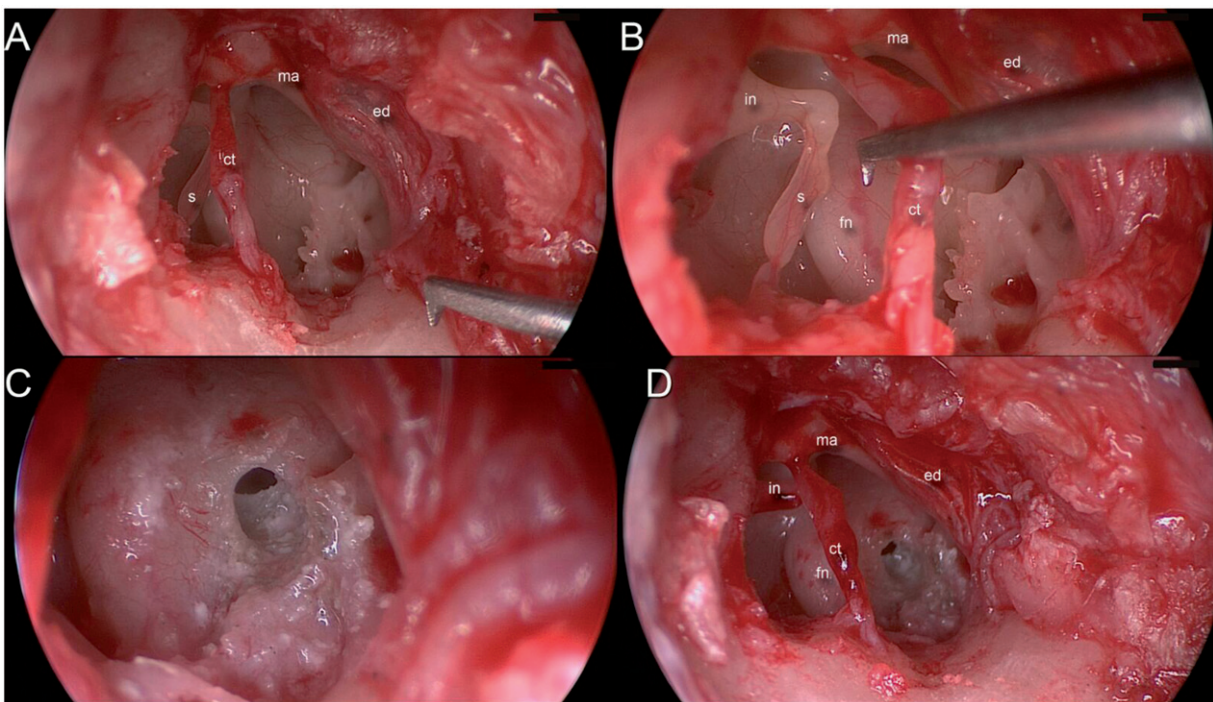
Figure 2. Endoscopic steps. Right ear. Panel A: the tympanometal flap is elevated, detecting the tympanic cavity. Panel B: the facial nerve is checked, in this case an anomalous course of the FN is found, running under anomalous stapes and obscuring the RW niche. Panel C: a promontorial cochleostomy was performed, maintaining the FN under endoscopic view. Panel D: final cavity after endoscopic steps. MA: malleus; ED: eardrum; S: stapes; IN: incus; CT: chorda tympani; FN: facial nerve.

periosteal flap was created and elevated, uncovering the mastoid bone. The skin of the posterior wall of the external auditory canal was elevated, and the tympanic cavity detected under microscopic view (Figure 3, Panel A). A mastoidectomy was performed until the antrum was exposed (Figure 3, Panel B); an anterior atticotomy was then performed uncovering the incudo-malleolar joint. The incus was removed, in order to create a wide connection between the posterior epitympanum and the mesotympanic spaces (Figure 3, Panel C). The receiver–stimulator of the implant was fixed and covered under the temporalis muscle. The array was gently pushed through the passage previously created, from the epitympanum into the mesotympanum; the array was then covered and gently inserted into the cochleostomy, through the external auditory canal (Figure 3, Panel D). A small piece of temporalis fascia was used with fibrin glue to seal the cochleostomy. The tympanomeatal flap was finally replaced and the external auditory canal packed with gelfoam. The subperiosteal flap was used to cover the receiver body and the retroauricular skin incision was sutured.

Results

Endoscopic findings

In all subjects, the endoscopic approach allowed to recognize the anatomical conformation of the tympanic cavity, the correct position of the FN and of the scala tympani, permitting a 'safe' cochleostomy under endoscopic view without injury of the FN. In all three patients, an anomalous course of the FN covering the RW niche was found, making difficult the surgical procedure on the RW membrane.

In Patients #1 and #3, the FN was dehiscent and located under the stapes over the promontory, obscuring the RW niche (Figure 2). The endoscopic approach was used to perform the cochleostomy over the promontory just anteriorly the RW. In Patient #2, a large erosion of the scutum and of the posterior wall of external auditory canal, with an extrusion of the array into the external canal, were found (Figure 4). The mastoid, the ossicular chain, the anomalous course of the tympanic segment of the FN and the array, fitted in a previous surgery in another hospital, were covered by cholesteatoma.
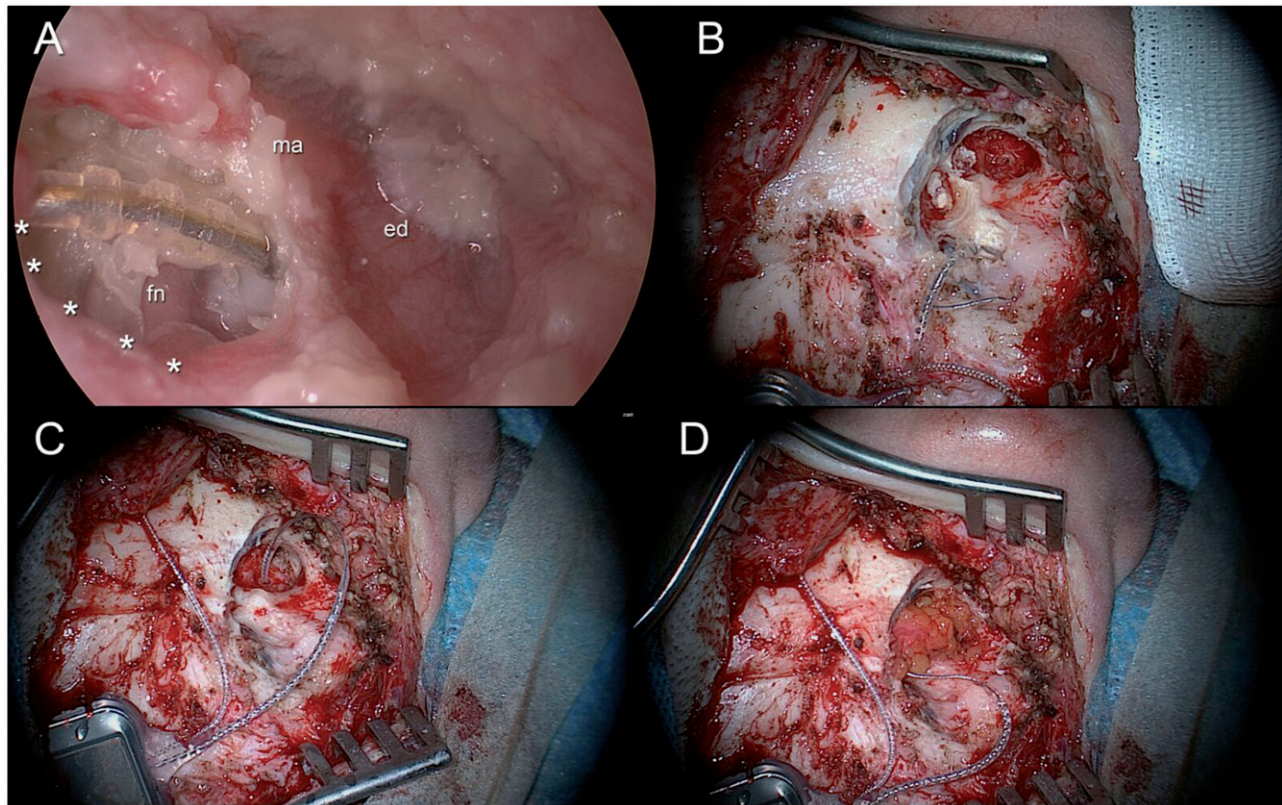
Figure 3. Microscopic steps. Right ear. Panel A: the mastoid and the external auditory canal are exposed under microscopic view. Panel B: a mastoidectomy is performed in a sclerotic mastoid until the antrum is visible. Panel C: an anterior atticotomy is performed and the incus removed, connecting the attic with the mesotympanum (white arrow), where the cochleostomy was previously performed. Panel D: the array is inserted through a transattic approach into the cochleostomy. EAC: external auditory canal; AN: antrum; SIS: sigmoid sinus; MCF: middle cranial fossa.
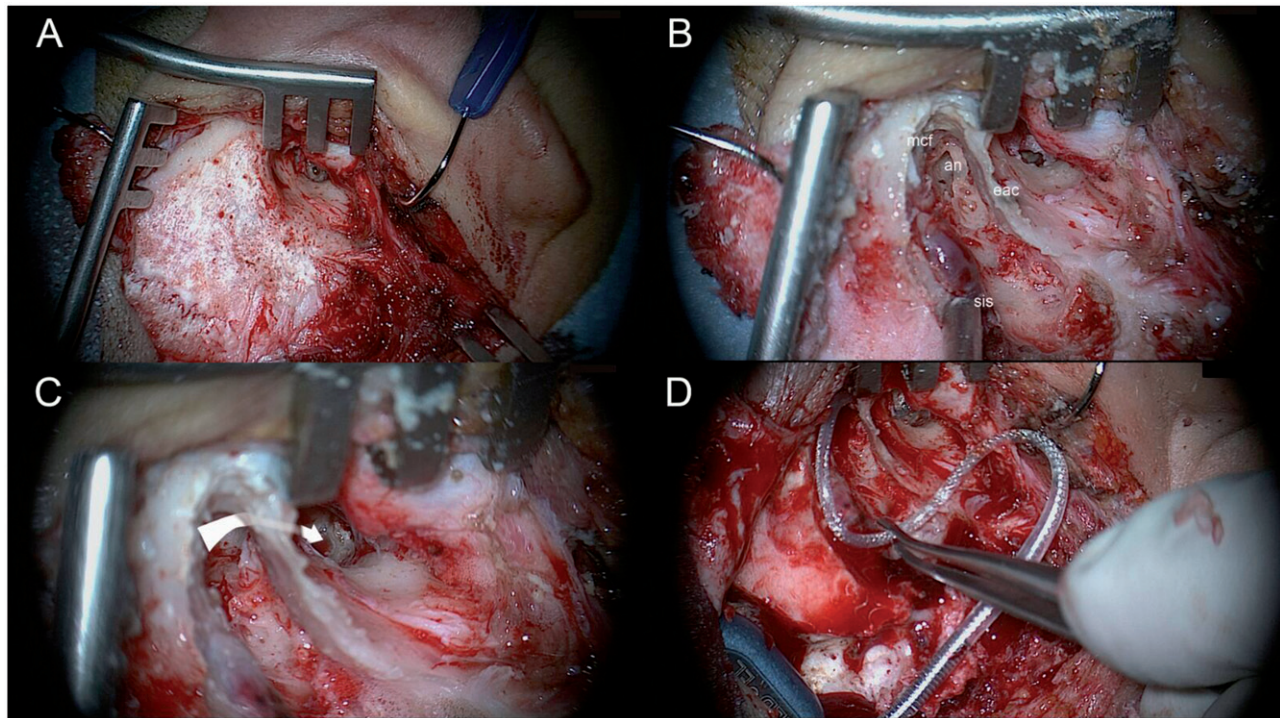
Figure 4. Patient #2 Panel A. Endoscopic view before surgery: a defect of the posterior wall of the external auditory canal is found (*); a cholesteatoma inside the tympanic cavity, completely covering the facial nerve and the array is also detected. Panel B. After endoscopic step the previous array was cutted, a mastoidectomy was performed in a sclerotic mastoid and the cholesteatoma was exposed; a large defect of the posterior wall of the external auditory canal was seen. Panel C. After subtotal petrosectomy the array was inserted into the cochleostomy previously created. Panel D. An obliteration of the surgical cavity was performed with fat. FN: facial nerve; ED: eardrum; MA: malleus.

The FN was located over the promontory. In this case, an endoscopic approach was used to remove the cholesteatoma from the tympanic cavity, maintaining the anomalous FN under endoscopic control. The previous implant was removed and an endoscopic approach was also used to perform the enlargement of the previous cochleostomy over the promontory.

Microscopic findings

In Patients #1 and # 3, due to the presence of sclerotic mastoid in both patients, the mastoid was removed until the antrum in the same manner as the transattic approach, then a posterior and anterior atticotomy was performed; the incus was removed creating a direct connection between the attic and the mesotympanic spaces, and the array was inserted via trans-attic approach into the cochleostomy previously created. In Patient #2, due to the extension of the cholesteatoma and the large defect of the posterior bony wall of the external auditory canal, a subtotal petrosectomy was performed. Neural responses were intraoperatively tested in all subjects and confirmed the correct insertion of the array.

Complications

All subjects were discharged from the hospital the day after surgery. No intraoperative or post-operative complications were observed in this series. In particular, no intraoperative gusher and no FN damage were found in any subject.

Audiological results

All patients exhibited profound hearing loss with no verbal production at preoperative evaluation. All of the implanted children showed a CAP score of 6 at the last follow-up (mean follow-up 12.5 months; range: 8–19 months).

Discussion

CHARGE syndrome has a prevalence of approximately 1/8500 newborns/year [7]. Children affected by CHARGE syndrome usually show abnormal cupped shaped pinna, hearing loss (conductive, sensorineural or mixed), abnormal ossicles and cochlear hypoplasia, absent semicircular canals (particularly lateral) [1], choanal atresia [8] and cranial nerve deficiencies,

including the vestibular, cochlear, facial and olfactory nerves [2]. Cochlea nerve aplasia and hypoplasia can occur *de novo* and are particularly associated with CHARGE syndrome [9,10]. Only a small proportion of children with CHARGE syndrome requires a CI, however conversely CHARGE syndrome is one of the more common syndromes seen in CI programs [2]. Several risks related with CI surgery in these patients should be considered. Parents counseling regarding risks and benefits of surgical procedure is mandatory, since our inability to predict audiological results and risks of the surgery must be stressed. First of all, the risk of FN injury during surgery is considerable, since the anomalous position of the FN into the tympanic cavity has been reported in 43% of patients with malformed cochlea [11], in association with sclerotic mastoid. Another important anatomical detail is related to the RW access; in some cases the FN is located over the RW niche, obscuring it. In these conditions, the identification of the RW region and the control of the FN course could be very difficult to obtain with the traditional microscopic CI surgery. Some authors suggested the use of the suprameatal approach route in order to perform a safe CI procedure and to reduce the incidence of FN injury in these patients [12]. Although from the previous experiences the selection of a specific surgical technique does not seem to influence the audiological outcome, the suprameatal access is valuable when important surgical landmarks (i.e. lateral semicircular canal and incus) are absent [12]. Also the inner ear distorted anatomy may represent a high risk during surgery. Birman et al. [2] reported 6 complications out of 10 children affected by CHARGE and requiring a CI surgery. In three children, the identification of the cochlea at the initial surgery was not possible. In one case the surgery was abandoned for difficulty in anatomical orientation of the inner ear and the procedure was repeated using a CT scan image guided surgery. In other two cases, an electrode misplacement occurred, due to difficult anatomical orientation. In our small series, the transcanal endoscopic approach allowed always to detect the anatomical landmarks into the tympanic cavity, as the position of the FN, the RW membrane and the scala tympani, in order to perform a 'safe' cochleostomy. In none of our patients, intraoperative and/or postoperative complications were observed and we were always able to obtain a proper and adequate insertion of the array in the cochlea, overcoming the suspension of the surgery, as reported in the literature [9]. No children developed facial palsy postoperatively, or had a twitching of the FN when the CI was activated,

responsible for abandoning the use of the implant [13]. In none of our patients meningitis or intraoperative cerebrospinal fluid (CSF) gusher, as it may be expected especially when an enlarged vestibular acqueduct or inner ear malformations are associated [9–14], have been reported. The endoscopic step also permits the transmastoid/transattic microscopic step to be performed more safely even in subjects with a sclerotic mastoid, since the surgeon knew the exact location of the anatomical structures into the tympanic cavity, most of all the course of the FN. Finally, this procedure allows to preserve the integrity of the posterior canal wall and to have a direct exposure of the tympanic cavity anatomy, avoiding the complex subtotal petrosectomy or an open approach as some authors have suggested for the treatment of malformed ears [15]. In our opinion, the use of endoscopic technique during CI surgery in subjects with normal anatomy of the ear is not indicated. The traditional approaches are well codified, with extremely rare complications and high success rate of treatment. In these cases, the transacanal approach should be avoided, according to the risks of this surgical procedure, such as iatrogenic cholesteatoma and the possible extrusion of the array. In our series, one subtotal petrosectomy was performed, since a large erosion of the posterior wall of the external auditory canal for an infiltrative cholesteatoma was present; nevertheless, this procedure is not necessary when a normal external auditory canal is found. The drawbacks of endoscopic CI consist mainly of the learning curve required for the surgeon to gain confidence with this one-handed technique that could be influenced by the presence of bleeding, so it is necessary to spend time performing a careful hemostasis. The final audiological outcome after implantation in this special group of patient is not predictable, since the distribution of neural tissue in inner ear anomalies is unstable. Our study suggests that verbal language development, even if not as high as in subjects with a normal anatomy and fitted with CI, is also possible for some children with CHARGE syndrome and congenital profound hearing loss. This is a preliminary report, since the small number of patients, and further studies must be done in order to understand benefits and limits of this surgical technique, but this preliminary experience indicates that the endoscopic-assisted surgical approach should be considered in subjects with malformed middle ear, in order to help the surgeon to have the right anatomical orientation and to perform a correct and safe insertion of the array.

Conclusions

On the basis of our personal experience, hereby illustrated, the use of transattical/endoscopic-assisted approach is highly suggested for cochlear implantation in patients with anatomical malformation of the ear, as in patients affected by CHARGE syndrome. Unlike the traditional microscopic approach, it allows better identification of middle ear structures. Of course, further studies will be necessary and long-term validation and lengthy follow-up are required, but the results obtained in this series of patients are very promising.

Disclosure statement

All of the authors have read and approved the manuscript. None of them have any financial relationship to disclose.

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