# Vestibular impairment in cochlear implanted children presenting enlarged vestibular aqueduct and enlarged endolymphatic sac

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## Introduction

The vestibular aqueduct is a bone canal that extends from the postero-medial part of the vestibule to the porus of the petrous bone. The enlarged vestibular aqueduct (EVA) was described by Valvassori and Clemis in 1978 by radiographic observation of 50 patients [1]. The endolymphatic sac (ES) is located on the posterior surface of the temporal bone, between the layers of dura mater. The EVA is the most common malformation of the inner ear and was described in 32% of pediatric patients with non-syndromic sensorineural hearing loss (SNHL) [2]. The most well-known cause of hearing loss associated to EVA is the mutation of the SLC26A4 gene (known as the *PDS* gene). Other associated abnormalities of the inner ear have been identified in 60% of cases (mainly an enlarged lateral semicircular canal and hypoplastic cochlea) [1].

Classically a vestibular aqueduct (VA) is considered abnormally enlarged if it is greater than 1.5 mm at midpoint on axial images. Other authors proposed newer criteria (width > 1 mm at the midpoint and/or an opening width > 1.9 mm at the operculum) [2-4]. EVA can also be associated with an enlarged endolymphatic duct and sac (EES), but it was rarely described in the literature [5]. According to Boston, EES was respectively found in 32% of children with non-syndromic SNHL [2]. Song et al. found bilateral EES in 55-94% of patients with EVA [6].

The clinical presentation includes fluctuating and progressive SNHL associated to varying degrees of vestibular impairment [2, 6-7]. Zalewski reported vestibular symptoms in nearly 50% in EVA patients [8]. In a cohort of 27 patients aged 3 to 12 years, Yang et al reported dizziness in 6 patients although 24 had abnormal vestibular tests [9]. In case of profound bilateral deafness bilateral cochlear implantation (CI) is an effective solution [10-12] but the intraoperative risk of oozing or gushing of cerebrospinal fluid (CSF) at the cochleostomy site

is high [13]. In 10 EVA children, Au and Gibson reported an initial spurt of perilymph during CI in 7 ears. Despite the potential interest of CI in EVA patients in the field of audiology [10, 13-14], only a few studies have investigated vestibular function after CI in these patients. In addition, few studies have evaluated the benefit of considering the size of the endolymphatic sac for CI [5].

The first objective of this study was to find out whether in EVA children candidates to CI, a higher endolymphatic sac volume was predictive for higher rates of post-surgical vestibular complications. Secondly, we analyzed the evolution of subjects with vestibular deficiencies appearing in the aftermath of CI over a 12-month period.

## Material and Method

*Population*

We retrospectively included children associating bilateral profound SNHL and EVA, and who benefited from cochlear implantation during the last 2 years. We included only the patients who benefited from a round window insertion, which was reported as a non-invasive technique preserving residual hearing [15] and is thought to limit the risk of vestibular impairment [16]. Thus, the risk of selecting patients with postsurgical vestibular impairment due to a possible traumatic insertion was limited. All have been implanted with electrodes designed for atraumatic insertion (from three manufacturer, see table 1). All children benefited from a vestibular assessment with the recording of cervical vestibular evoked myogenic potentials (cVEMPs) and video head impulse test (VHIT, Ulmer, France) for each semi-circular canal before CI and 6 months after surgery. All subjects underwent HRCT of the petrous bone and, labyrinthine MRI with T2 Drive sequences (Philips Ingenia 3T MRI, Philips healthcare, Amsterdam, Nederland) at least 6 months before surgery. The imagery protocol used in our Cochlear Implantation Center is described below.

The investigation adhered to the principles of the Declaration of Helsinki. Written informed consent was obtained from the children's parents.

*Radiological assessment*

For high resolution cranial tomodensitometry (HRCT), the tomodensitometric diagnostic criteria for EVA proposed by Valvassori and Clemis were used [1]. The VA was considered enlarged according to Valvassori criteria (>1.5 mm at midpoint of the VA on axial images) and the newer Cincinatti criteria (width > 1 mm at the midpoint in the coronal plane and/or an opening width >1,9 mm at the operculum) [1-2].

3T MRI of the petrous bone and inner ear structures was performed in all patients before CI (Philips Ingenia 3T MRI, Philips healthcare Amsterdam, Nederland). Labyrinthine 3DT2 high-resolution DRIVE (DRIVen Equilibrium pulse) sequences were used to obtain a volumetric acquisition of the ES (Philips Portal, Philips healthcare Amsterdam, Nederland). Endolymphatic volume comprised vestibular aqueduct and ES. It was measured from the posterior part of the vestibule to the end of the lateral part of the ES (**Figure 2**).

*Vestibular assessment*

The neurotological examination as well as the vestibular assessment were carried out by the same clinician in all subjects. Vestibulo-Ocular Reflex (VOR) was assessed in each subject before and 6 months after CI. The Ulmer VHIT system (Ulmer, Marseille, France) was used to measure the gain of the VOR for each of the six semicircular canals using 10 validated head turns (up to 10 Hz stimulation) in the plane of each SCC. Otolithic function was assessed before and 6 months after surgery by cVEMPs elicited in bone conduction (BC) as previously described for children [17]. Although the utricle may also respond to BC stimuli, the presence of cVEMPs indicates predominantly human saccular response function [18]. Although cVEMPs were studied in BC, otoscopy and tympanometry were verified in each subject before cVEMPs assessment. Children either lay on a clinical table or sat on their parent's lap. A Collin company bone vibrator (Collin medical, France) was applied on the mastoid to test the sacculo-spinal reflex in BC. Brief acoustic clicks (100 µs) were delivered unilaterally at a frequency of 500 kHz and 70 dB SPL. During this stimulation, the activity of the sternocleidomastoid muscle was recorded by electromyography (EMG). If the cVEMPs response was absent or inferior to 50µV, the response was marked “absent”.

*Groups constitution*

Two groups were constituted according to the presence or not of a vestibular impairment 6 months after CI. The presence of an intervention-related vestibular deficit was defined by a decrease in the VOR gain on the VHIT test on one of the semicircular canals by 0.2 or more and/or a loss of cVEMPs. Hence, 6 children were in the impaired group, and 9 in the non-impaired group. In each group, walking was acquired for each child and there was no delay in motor development. All children had a normal vestibular assessment before CI, except 2 in the impaired group who had horizontal canal impairment.

*Statistical analysis*

Data were entered into Microsoft Excel (Microsoft Corp., Redmond, WA) spreadsheet and the Sigma Stat software (Systat Software Inc., San José, USA) for further analysis. In both groups, the volume of the ES was compared. Nonparametric Mann-Whitney U test was used as law of normality was not respected. All results were expressed as the mean ± standard deviation (SD). Findings were considered statistically significant when the P value was less than 0.05.

## Results

*Global results*

Fifteen patients (8 males, 7 females) completed all inclusion criteria and were selected (table 1). Among the 15 patients, the average age was 40 months (range 20 to 72). CI was performed on the right side in 6 patients and on the left side in 9 patients. Pre and postoperative cVEMPs thresholds, VHIT gains for each canal and MRI measures of endolymphatic volume are presented in **table 1**.

| Patient | Age at CI (M) | Ear of CI | VHIT LC/AC/PC (Gain) Before CI | VHIT LC/AC/PC (Gain) After CI | cVEMPs Threshold (dB) Before CI | cVEMPs Threshold (dB) After CI | Per op findings | MRI ES volume (CI ear, cm3) | Associated inner ear malformation (etiology) | Clinical post op findings | CI manufac electrode a |
|---|---|---|---|---|---|---|---|---|---|---|---|
| **Non impaired group** | | | | | | | | | | | |
| 1 | 48 | LE | 0.9/0.7/0.8 | 0.8/0.8/0.7 | 85 | 95 | N | 0.29 | Mondini | Nausea | Cochlear C slim straigh |
| 2 | 46 | LE | 1/0.9/0.9 | 1/0.9/0.8 | 85 | 85 | Oozing | 0.05 | No | 0 | Advanced HR 90K Sl |
| 3 | 42 | LE | 0.9/0.7/0.8 | 0.9/0.7/0.8 | 85 | 85 | Oozing | 0.09 | Mondini | 0 | Cochlear C slim straigh |
| 4 | 20 | LE | 0.9/0.7/0.7 | 0.9/0.7/0.7 | 80 | 80 | Oozing | 0.13 | Mondini | 0 | Cochlear C slim straigh |
| 5 | 40 | RE | 0.8/0.7/0.7 | 0.8/0.6/0.7 | 85 | 85 | Oozing | 0.11 | No | 0 | Cochlear C slim straigh |
| 6 | 34 | RE | 0.9/0.7/0.7 | 0.9/0.7/0.7 | 90 | 90 | Oozing | 0.13 | No | 0 | Advanced HR 90K Sl |
| 7 | 40 | RE | 1/0.8/0.8 | 0.9/0.7/0.7 | 90 | 95 | N | 0.09 | No | Nausea | Medel Syn flex24 |
| 8 | 24 | RE | 0.9/0.8/0.8 | 0.9/0.7/0.7 | 85 | 80 | N | 0.04 | No | GI | Medel Syn flex24 |
| 9 | 35 | LE | 0.9/0.8/0.7 | 0.8/0.7/0.7 | 85 | 90 | Oozing | 0.1 | No | Nausea | Advanced HR 90K Sl |
| **Impaired group** | | | | | | | | | | | |
| 10 | 24 | RE | 1/0.8/0.8 | 0.4/0.1/0.1 | 90 | Absent | Oozing | 0.70 | No | Nausea/ GI | Advanced HR ultra 3I |

| | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | | | | | | | SlimJ |
| 11 | 72 | LE | 0.6/0.7/0.7 | 0/0.6/0 | 95 | Absent | Oozing | 0.42 | No (Pendred) | GI | Cochlear C slim straigh |
| 12 | 28 | RE | 0.5/0.7/0.7 | 0.5/0.7/0.7 | 95 | Absent | Gusher | 0.08 | LC malformation | 0 | Medel Syn flex24 |
| 13 | 48 | LE | 0.8/0.7/0.7 | 0.5/0.5/0.1 | 90 | Absent | Gusher | 0.6 | No | Nausea/ GI | Cochlear C slim straigh |
| 14 | 28 | LE | 1/0.8/0.9 | 0.5/0.6/0.5 | 95 | Absent | Oozing | 0.26 | No (Pendred) | Nausea/ GI | Cochlear C slim straigh |
| 15 | 68 | LE | 0.8/0.8/0.7 | 0.6/0.5/0.5 | 85 | Absent | Gusher | 0.37 | Enlarged IAC | Nausea/ GI | Cochlear C slim straigh |

Table 1: Patient characteristics with the measurements performed for the VHIT test and the cVEMPs in BC before and after CI, with measures of the endolymphatic volume (vestibular aqueduct and the ES) in MRI. *CI: cochlear implant. LC: lateral semicircular canal, AC: anterior semi-circular canal, PC: posterior semi-circular canal, cVEMPs: cervical vestibular evoked myogenic potentials, VHIT: Video head impulse test, ES: endolymphatic sac, IAC: Internal auditory canal, N: Normal. GI: Gait instability.*

Cochlear dysplasia ("Mondini" malformation) was the most common temporal bone anomaly and was found in 3 patients. During cochlear implant electrode insertion, a gusher was only found in 3 patients, an oozing was found in 9 patients. In the non-impaired group, an oozing was observed in 6 of 9 patients during CI insertion (no gusher was observed). In the impaired group, a gusher was observed in 3 out of 6 children and an oozing in the 3 other children. The mean endolymph volume was significantly higher in the impaired group (0.40 cm3 ± 0.23, range 0.08 to 0.70) than in the non-impaired group (0.11 cm3 ± 0.07, range 0.04 to 0.29; p=0.029). In the impaired group, cVEMPs were absent after cochlear implantation in all children whereas a canalar impairment (assessed by VHIT) was only found in 5 out of 6 children. If we consider only postoperative VHIT results, the mean volume was significantly higher in the impaired group (0.47 cm3 ± 0.18, range 0.26 to 0.7) than in the non-impaired

group (0.11 cm3 ± 0.07, range 0.04 to 0.29; p=0.004). The results were similar for other semi-circular canals ipsilateral to the implanted ear because if a vestibular impairment was observed on one SSC, all semi-circular canals were impaired.

*Evolution of vestibular function in group 2 after 1-year*

Four children of the impaired group were followed during one year with recurrent vestibular assessment (at 1 month, 6 month, and 12 month). Data are available in **Table 2**. All children had walking difficulties or nausea in the days following CI that is why they benefited from specialized physiotherapy for a period of 6 to 12 months. At the end of vestibular rehabilitation, all children recovered a lateral canal function (i.e. an improvement in VHIT gain) and a saccular function (i.e. cVEMPs reappeared).

| | **Clinical post op findings** | **VHIT gain for LC** | | | | **cVEMPs threshold (dB)** | | |
|---|---|---|---|---|---|---|---|---|
| **Patient** | | Before CI | One month after CI | Six months after CI | One year after CI | Pre CI | Post CI | At 1 year |
| 10 | Nausea/GI | 1 | 0.1 | 0.4 | 0.6 | 90 | Absent | 110 |
| 13 | Nausea/GI | 0.8 | 0.2 | 0.5 | 0.7 | 90 | Absent | 95 |
| 14 | Nausea/GI | 1 | 0.2 | 0.5 | 1 | 95 | Absent | 110 |

| | | | | | | | | |
|---|---|---|---|---|---|---|---|---|
| 15 | Nausea/GI | 0.8 | 0.5 | 0.6 | 0.6 | 85 | Absent | 90 |

Table 2: Vestibular follow-up (cVEMPs and VHIT gain for LC) one year after CI in the impaired group. *CI: cochlear implant. LC: lateral semicircular canal. GI: Gait instability.*

*Case report*

A 2-year-old child with a history of profound bilateral deafness was assessed in our unit before CI. Walking was acquired at 14 months. HRCT showed large vestibular aqueducts without Mondini dysplasia; MRI in a T2-weighted axial plane showed bilateral enlarged ES that was not predictable on HRCT (**Figure 1 and 2**).

Vestibular assessment before IC was normal with cVEMPs showing a bilateral 95 dB threshold and VHIT gain of 0.8, 0.7 and 0.6 for lateral, anterior and posterior semi-circular canals respectively. Pendred syndrome was suspected because there was a mutation in the PDS gene. Left ear was chosen for first CI on audiological criteria.

During surgery, oozing but no gusher, cerebro-spinal fluid leak was controlled with a muscle plug. Early after implantation (at day 1), the child was annoyed by nausea with vomiting and the parents reports falls during a period of 2 or 3 days; one week after CI, clinical assessment showed positive Halmagyi sign on the left (implanted) side with skew deviation, ocular torsion, head tilt toward the left eye, evocative of ocular tilt reaction [19]. CVEMPs were only present at 95 dB on the right side; VHIT gains were found at 0.2, 0.5 and 0.1 for left lateral, anterior and semi-circular canals respectively. Vestibular physiotherapy quickly improved the

child after CI. At 6 months-follow-up, the VHIT gain for the left lateral canal was improved to 0.5. Gains for anterior and posterior canals were respectively 0.7 and 0.5. One year later, the gain was almost 1 (with covert saccades) for lateral and anterior canals but 0.4 for the posterior canal. At this time, cVEMPs were found unchanged at 110 dB threshold on the left side. On the right side, the assessment remained unchanged and normal. As left vestibular impairment improved, right CI was scheduled 14 months after the left CI.

## Discussion

Our main objective was to highlight the risk of vestibular impairment following CI in children candidates presenting EVA. We found that the mean endolymph volume of VA and ES was significantly higher in the group with vestibular impairment after CI than in the non-vestibular impaired group. Our data suggests that children with EVA and EES were at greater risk of vestibular loss during CI. According to Cushing et al. 50% of children with profound bilateral SNHL (with no EVA) have some abnormality of vestibular end-organ function [20]. However, in our study 13 children out of 15 did not present a vestibular impairment before CI. It is to note that documentation of pre- and post-operative vestibular function surrounding pediatric cochlear implantation are limited. According to Yong et al. it could be reflective of the fact that many centers do not routinely perform such testing [21]. In their recent meta-analyses, it is suggested that there is a significant risk for abnormal VEMP responses in pediatric patients undergoing CI, with a significant relative risk of 1.8. Concerning VHIT testing, there is a lack of available data in implanted non EVA children [21] but in adult population, canalar function as studied with VHIT is usually little affected [16,22]. Our study has some limitation given the small size of samples but in another hand it is quite difficult to dispose of complete

vestibular assessment in young children before and after cochlear implantation especially in this rare pathology. It should be noted that videonystagmography (VNG) with caloric testing is performed whenever possible but was only possible in a few children in this study, for this reason it was not considered. In addition, two children in the impaired group previously had reduced gain on the horizontal channel only (0.6 and 0.5) on the VHIT test. The saccular function was preserved. These children presented a degradation of gain in all channels at VHIT and a loss of cVEMPs after implantation. We therefore chose to include them in the impaired group.

In our study, during surgery, an oozing or a gusher was found in 12 patients with EVA. However, the sample size of the study is too small to conclude that a gusher is more frequently present at higher endolymphatic volumes. Kim et al showed previously that in EVA patients the size of the ES was not correlated with CSF gushers [5].

Secondarily, in 4 out of 6 children in the impaired group, the same data has been collected during a 1-year follow-up. All of these 4 children benefited shortly after surgery from specialized physiotherapy for a period of 6 to 12 months. On this one-year period, they regained progressively a satisfactory vestibular function. We found no studies in the literature concerning the evolution of the vestibular balance following cochlear implantation in EVA patients. The small size of our sample size does not allow us to reassure on the clinical evolution of these patients.

Only a few studies in literature focused on vestibular assessment in EVA children. Song et al also studied the correlations between vestibular symptoms and the size of the VA but also the degree of hearing loss [8]. Twenty-two EVA patients were included (median age 8 years) and besides an auditory assessment, they performed a vestibular assessment: VNG with bi-thermal caloric and vibration, head impulse test. They found no correlation between the width of the

VA and hearing thresholds, and no correlation between mean hearing loss and vestibulopathy. Other authors found a positive correlation between VA width and progressive sensorineural hearing loss [2] and a significant correlation between hearing threshold at a low frequency and vestibular function in patients with bi-allelic SLC26a4 mutations [7]. Some may argue that the volume of the endolymphatic sac could vary with age or even over time, but to the best of our knowledge only few studies have been conducted on this subject [23].

We raised the interest of the association of HRCT and MRI to better evaluate the risk of vestibular impairment after cochlear implantation in EVA children. The idea of associate tomodensitometry and MRI was already described in the literature [9,24]. Okamoto et al. underlined the fact that in some patients with SNHL and EES, the vestibular aqueduct may not appear dilated on tomodensitometry [24]. Therefore, we think that MRI is necessary for a correct diagnosis of EES by showing the entire volume of the endolymphatic drainage system (especially the extra bony part of the endolymphatic sac, not visible in HRCT). According to Okamoto et al, EVA should more correctly be termed “large endolymphatic duct and sac syndrome”, and prominent EES might predict poor prognosis in this syndrome [24]. The mechanism of hearing loss in EVA is still unclear. Sudden changes in CSF pressure may induce reflux of hyperosmolar contents in the ES in the cochlear duct, or vestibular hair cell destruction due to osmotic and chemical imbalance [8,24]. Pressure gradients may have a greater effect on the auditory system, but EVA could also act as a third window on the scala vestibuli side leading to conductive hearing loss.

Thin-slice MRI, besides providing a clear assessment of cochlear nerve integrity, central nervous system abnormalities allow evaluating soft tissues and fluids of the endolymphatic duct and sac directly [25]. To evaluate the correlation between SNHL and EVA, Campbell et al proposed an endolymphatic duct measure on 1.5 or 3T MRI, with qualitative assessments (heterogeneity) of the signal inside the ES [25]. They use four measures: endolymphatic duct

width closest to the vestibule, at the midpoint between the vestibule and ES, ES length, and ES width. They found that endolymphatic duct width measured near its origin at the vestibule and the presence of ES signal heterogeneity correlated with worse hearing levels [25]. We used a method of measuring the total volume of the ES, from the vestibule to its termination at the exit of the petrous bone. We did not study in detail the signal density of the ES, but an endolymphatic sac tumor (ELST) was systematically ruled out. In a recent review of literature, ELST were described as rare lesions of the petrous temporal bone originating from the epithelium of endolymphatic duct and ES. The radiological aspect was recently reviewed by Le et al in a study of 14 adults [26]. HRCT findings revealed spiculated, stippled, or reticular high density within the tumors. MRI showed patchy and or speckled hyper intensity on unenhanced T1.

## Conclusion

Enlarged ES in enlarged vestibular aqueduct children could increase the risk of severe post CI vestibular impairment. The small sample size of the study does not allow for robust conclusions and further studies need to be conducted to validate these findings. However, to minimize this risk prior CI surgery - besides standard HRCT temporal bone assessment - MRI measurement of the ES volume should be systematically performed. Although a "spontaneous" (but poor) vestibular compensation is possible, the postural control in these subjects could be improved after CI surgery by intensive specialized physical therapy. To avoid severe vestibular ataxia by bi-vestibular post CI surgery failure, sequential cochlear implantation should be systematically recommended especially in all patients with EVA associated to EES.

**Legends**

Figure 1: A: High resolution cranial tomodensitometry showing the enlarged vestibular aqueduct. B and C: 3T-MRI with T2 DRIVE images in axial (B) and sagittal (C) plane showing the bilateral enlarged endolymphatic sac.

Figure 2: A: MRI measures of right endolymphatic sack volume. B: MRI reconstruction images of enlarged endolymphatic sacks and ducts in axial plane. *ES: endolymphatic sack, SCC: semicircular canals, VA: vestibular ~~a~~queduct~~.~~*

Figure 3: Measurement of vestibulo-ocular reflex gains at high frequencies at the Video Head Impulse Test (VHIT) for each semicircular canal, before, after IC, 6 and 12 months after CI in the patient no 10.

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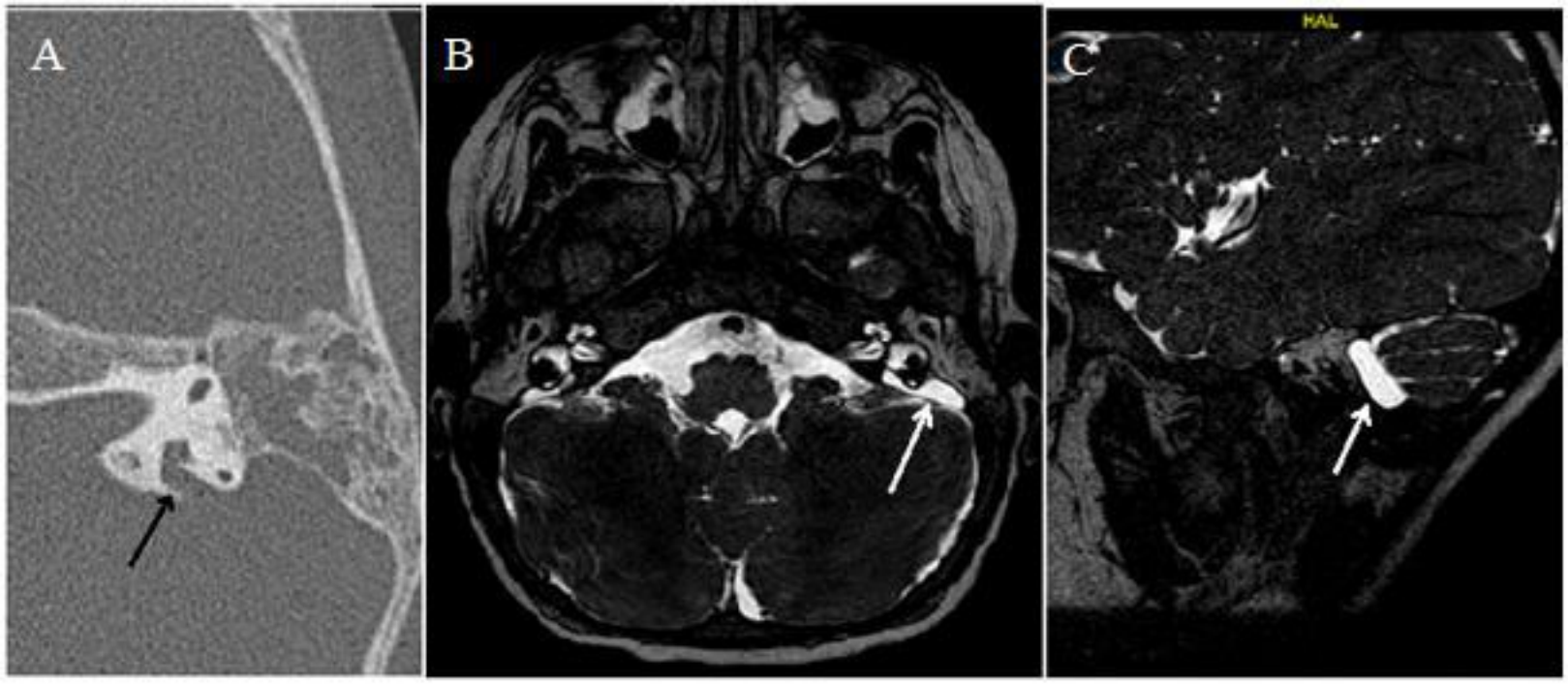
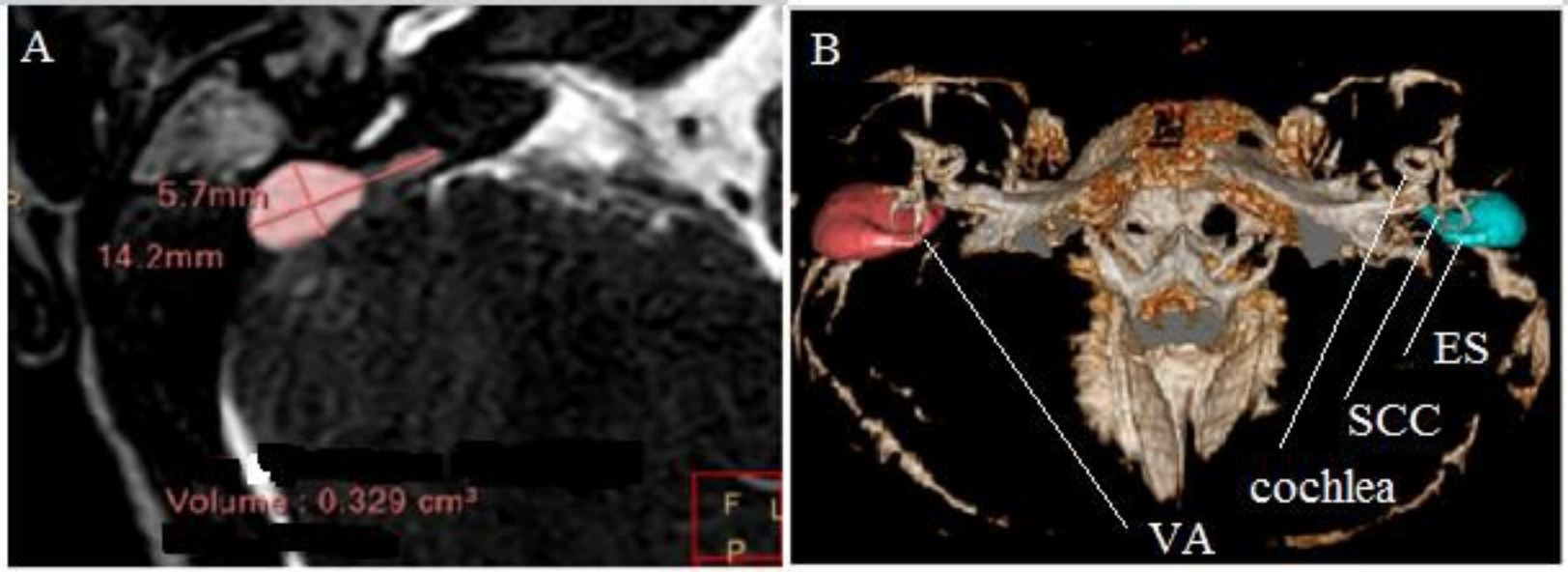
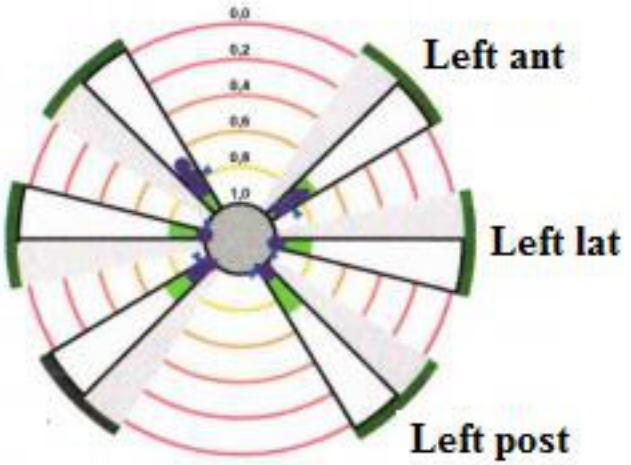
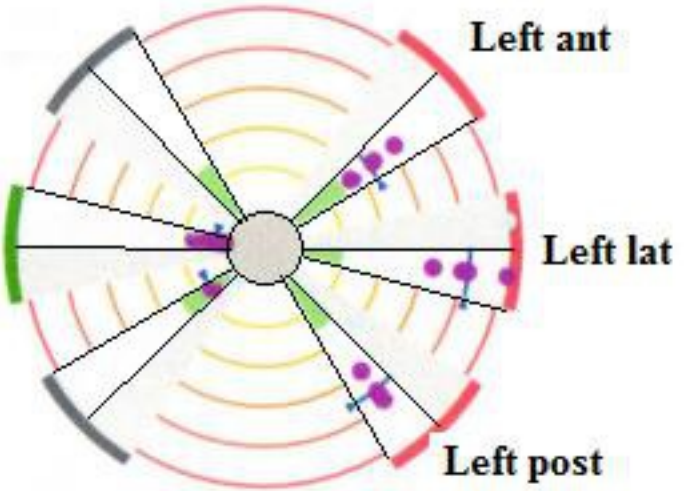

A
B
C

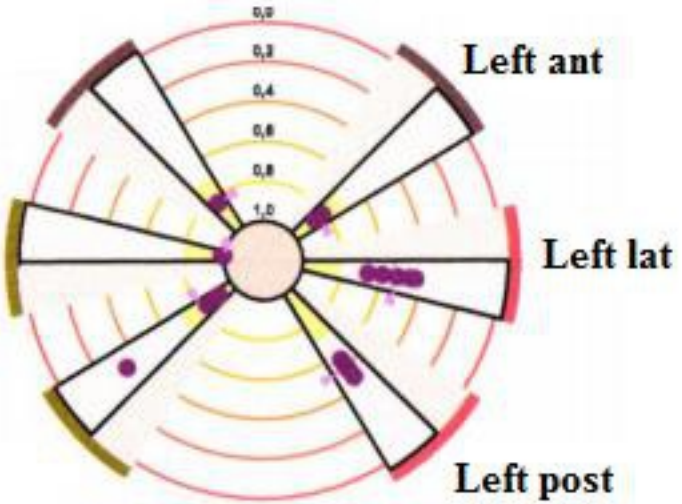

A
5.7mm
14.2mm
Volume : 0.329 cm³
F L
P
B
ES
SCC
cochlea
VA



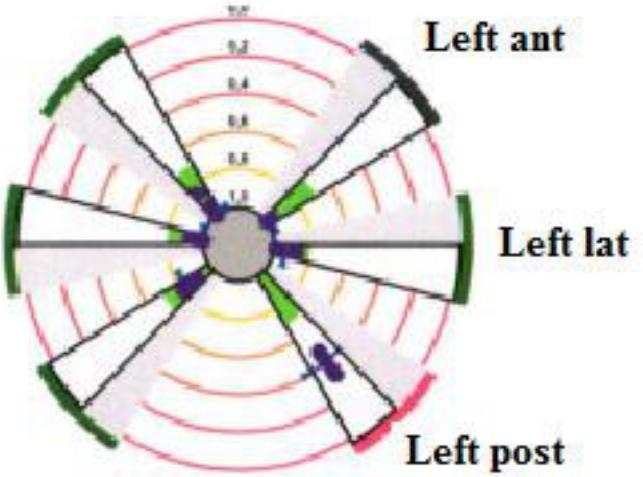

Before CI




After left CI




6 months after left CI




12 months after left CI