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OTOLOGY

# Cochlear implantation in patients with inner ear bone malformations with posterior labyrinth involvement: an exploratory study

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**Abstract** Inner ear bone malformations are one cause of profound sensorineural hearing loss. This investigation focused on those affecting the posterior labyrinth, especially enlarged vestibular aqueduct syndrome, which is associated with fluctuating and progressive hearing loss. The objectives of this study were to analyze the behavior of the electrical stimulation, auditory functionality and linguistic development in patients with inner ear malformations involving the posterior labyrinth. The study included ten patients undergoing cochlear implantation (cases: five with enlarged vestibular aqueduct, two with vestibular aqueduct stenosis/aplasia, and three with semicircular canal disorders). Post-implantation, data were gathered on the electrical stimulation threshold and maximum comfort levels and on the number of functioning electrodes. Evaluation of Auditory Responses to Speech (EARS) subtests were used to assess auditory functionality and language acquisition at 6, 12, and 24 months post-implantation. Results were compared with findings in a control group of 28 cochlear implantation patients without these malformations. No significant differences were found between case and control groups in electrical stimulation parameters; auditory functionality subtest scores were lower in cases than controls, although the difference was only statistically significant for some subtests. In conclusion, cochlear implantation patients with posterior labyrinth bone malformations and profound hearing loss, including those with enlarged vestibular aqueduct syndrome, showed no significant difference in electrical stimulation threshold with controls. Although some auditory functionality test results were lower in cases than in controls, cochlear implantation appears to be beneficial for all patients with these malformations.

**Keywords** Hearing loss · Enlarged vestibular aqueduct syndrome · Cochlear implantation · Posterior labyrinth bone malformations

**Abbreviations**

| | |
|---|---|
| CHARGE | Coloboma, heart defects, choanal atresia, growth retardation, genital hypoplasia and ear anomalies |
| EVAS | Enlarged vestibular aqueduct syndrome |
| LIP-Profile | Listening progress profile |
| MTP | Monosyllabic-trochee-polysyllabic-word test |
| OLD | Open list disyllabics (open-set test) |
| CLD | Closed list disyllabics (closed set test) |
| CT | Computed tomography |
| THR | Threshold of electrical stimulation |
| MCL | Maximum comfort level |
| μC | Microcoulomb |
| EARS | Evaluation of auditory responses to speech |

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## Introduction

Inner ear bone malformation is one cause of profound hearing loss [1, 2]. Malformations that affect the posterior labyrinth include the enlarged vestibular aqueduct syndrome (EVAS), first described by Valvassori [3, 4]. The percentage of inner malformations represented by EVAS ranges from 7 to 32 % [5–9], and it is one of the most frequent diagnoses in children with profound sensorineural hearing loss [6–8], the mechanism of such loss is unclear, and there are no established criteria for diagnosis [7]. EVAS is a congenital malformation, often begins in childhood and subsequently progresses to profound hearing loss [5, 10], generally postlingual [11]. This anomaly should not produce auditory deficit, because the cochlea of these patients appears undamaged. Nevertheless, it is frequently associated with fluctuating or progressive hearing loss, occasionally triggered by a minor traumatic brain injury (TBI) and sometimes accompanied by vertiginous syndrome [9]. A vestibular aqueduct is considered to be enlarged when its diameter is >2 mm [5, 9], although Winbrad et al. reported that the normal vestibular conduct diameter ranges between 0.4 and 1 mm [10]. Others reported that anatomical alterations of the vestibular aqueduct include aplasia or total stenosis [2, 12], and malformations of the posterior labyrinth can also affect the semicircular canals, producing their aplasia, hypoplasia, or stenosis [12]. If these conditions progress to profound hearing loss, the final treatment is cochlear implantation [13], with the risk of perilymphatic gusher during surgery, especially in patients with aqueduct enlargement [7, 9, 14].

The main objective of this study was to analyze the behavior of electrical stimulation parameters during cochlear implant programming in patients with inner ear malformations involving the posterior labyrinth. A further objective was to assess the auditory functionality and linguistic development of these patients. Cochlear implantation in children with EVAS or others posterior labyrinth malformations is challenging in the audiological aspect and it is little investigated in the literature. In recent studies, good audiological and speech results have been obtained after cochlear implantation in these patients when compared with patients without malformations [5–10, 12, 15].

## Materials and methods

A case–control, observational, retrospective study was undertaken in consecutive patients with inner ear bone malformation involving the posterior labyrinth who received a cochlear implant between 2006 and 2011 in San Cecilio University Hospital, Granada (Spain). No exclusion criteria were applied, and the sample included all 10 patients; the mean age at implantation was 14.7 years (SD 15.20) and the median age was 4 years (Table 1). For this study, ethical approval was obtained by the ethical committee of the San Cecilio University Hospital and Faculty of Medicine, Granada.

Before cochlear implantation, all patients underwent helical high-resolution computerized tomography (CT) in slices of 0.5–1 mm for diagnostic and morphologic examination of the inner ear [13] and magnetic resonance imaging (MRI) to examine the membranous labyrinth and cochlear nerve [15]. The following posterior labyrinth malformations were detected: EVAS ($n = 5$, 50 %), vestibular aqueduct stenosis/aplasia ($n = 2$, 20 %), and semicircular canal alterations ($n = 3$, 30 %). None of these patients had inner ear bone malformation involving the anterior labyrinth. Table 2 summarizes the characteristics of the patients (cases).

A group of 28 cochlear implant patients (controls) with no inner ear anomalies was included in the study; their distribution by sex and age at implantation is reported in Table 1. These patients have not submitted any associated syndrome.

The following cochlear implants were used: Med-el Pulsar CI100 (Innsbruck, Austria) ($n = 4$), Med-el Sonata Ti100 (Innsbruck, Austria) ($n = 2$), Advance Bionics Hires 90k (Laubisrütistrasse, Switzerland) ($n = 3$) and Cochlear Nucleus (Macquarie University, Australia) ($n = 1$). The selection of implant depended on the availability and was not related to the characteristics of the patient. In our patients, different electrode arrays (short, split…) were not used. After cochlear implantation and its activation and programming, data were gathered on the electrical stimulation threshold (THR) and maximum comfort level MCL (both measures in microcoulombs) and on the number of active electrodes. Auditory functionality scores (percentage 0–100 % of correct answers) were obtained at 6, 12, and 24 months in the following subtests: Listening Progress (LIP) profile, Monosyllabic-Trochee-Polysyllabic (MTP) 3-word test, MTP 6-word test, MTP 12-word test, closed list disyllabics (CLD) and open list disyllabics (OLD) from the Evaluation of Auditory Responses to Speech (EARS), [16, 17] battery developed by the Department of Clinical Research of MEDEL (Innsbruck, Austria).

In the statistical analysis no parametric test was used due to the sample size ($n < 30$). The Chi-square test was used to compare proportions between groups (cases and controls) and when it did not meet the conditions of validity, Fisher's exact test was applied.

Analysis of differences was done by Mann–whitney *U* test (to analyze the differences between the mean values of quantitative variables between two groups of study) and

**Table 1** Distribution of patients in case and control groups

| | Patients with malformation (cases) | Patients without malformation (controls) |
|---|---|---|
| Number of patients | 10 | 28 |
| Sex | 6 males (60 %) | 17 males (60.7 %) |
| | 4 females (40 %) | 11 females (39.3 %) |
| Age at implantation (mean) | 14.7 years (SD 15.20) | 9.68 years (SD 13.72) |
| Age (median) | 4 years | 2.5 years |

*Malformation* inner ear bone malformation with posterior labyrinth involvement

**Table 2** Characteristics of patients with posterior labyrinth malformation who underwent cochlear implantation

| Patient | Sex | Age at implantation (years) | Type of malformation | Type of hearing loss | Degree of hearing loss (dB) | Personal history | Surgical complications |
|---|---|---|---|---|---|---|---|
| 1 | Male | 2 | EVAS | Prelingual | 90 | Pendred syndrome | – |
| 2 | Female | 42 | EVAS | Postlingual (Progressive from childhood) | 90 | – | Gusher (during the intervention) |
| 3 | Male | 5 | EVAS | Perilingual | 100 | Fever seizures with convulsions | – |
| 4 | Female | 13 | EVAS | Postlingual (Progressive from childhood) | 95 | – | Gusher (during the intervention) |
| 5 | Female | 55 | EVAS | Postlingual (progressive from childhood) | 100 | – | – |
| 6 | Male | 4 | Vestibular aqueduct aplasia | Prelingual | 100 | Neonatal sepsis | – |
| 7 | Male | 3 | Vestibular aqueduct stenosis | Prelingual | 100 | – | – |
| 8 | Male | 4 | Semicircular canal aplasia | Prelingual | 95 | Mild psychomotor retardation | – |
| 9 | Male | 16 | Semicircular canal stenosis | Prelingual | 90 | TBI at the age of 2, progressive hearing loss | – |
| 10 | Female | 3 | Semicircular canal stenosis | Prelingual | 100 | – | – |

*EVAS* enlarged vestibular aqueduct syndrome

Kruskal–Wallis test (was used to compare mean differences in more than two groups).

## Results

The behavior of electrical stimulation parameters, auditory functionality and linguistic development are presented below. Figure 1 exhibits the stimulation THR values obtained in the programming procedure. No significant differences were found between cases and controls or between the subgroup of patients with EVAS and controls in THR, MCL, or number of functioning electrodes ($p > 0.05$ in all cases).

Table 3 displays the auditory perception test results. Significant differences ($p < 0.05$) were found between cases and controls in the MTP 6-word test at 6, 12, and 24 months post-implantation, in the MTP 12-word test at 24 months, and in the OLD at 12 months. No significant differences were found in the LIP, MTP 3-word test, or CLD at 6, 12, or 24 months or in the OLD at 24 months.

## Discussion

Profound hearing loss is the most frequent sensorineural disorder in newborns [2]. The reported frequency of inner ear bones malformations among patients with profound hearing loss ranges between 7 and 20 % in different studies using imaging techniques [1–5]. The present study only includes 10 cases of inner ear malformation with posterior labyrinth involvement, including 5 patients with EVAS, 2 with vestibular aqueduct stenosis/aplasia stenosis and 3

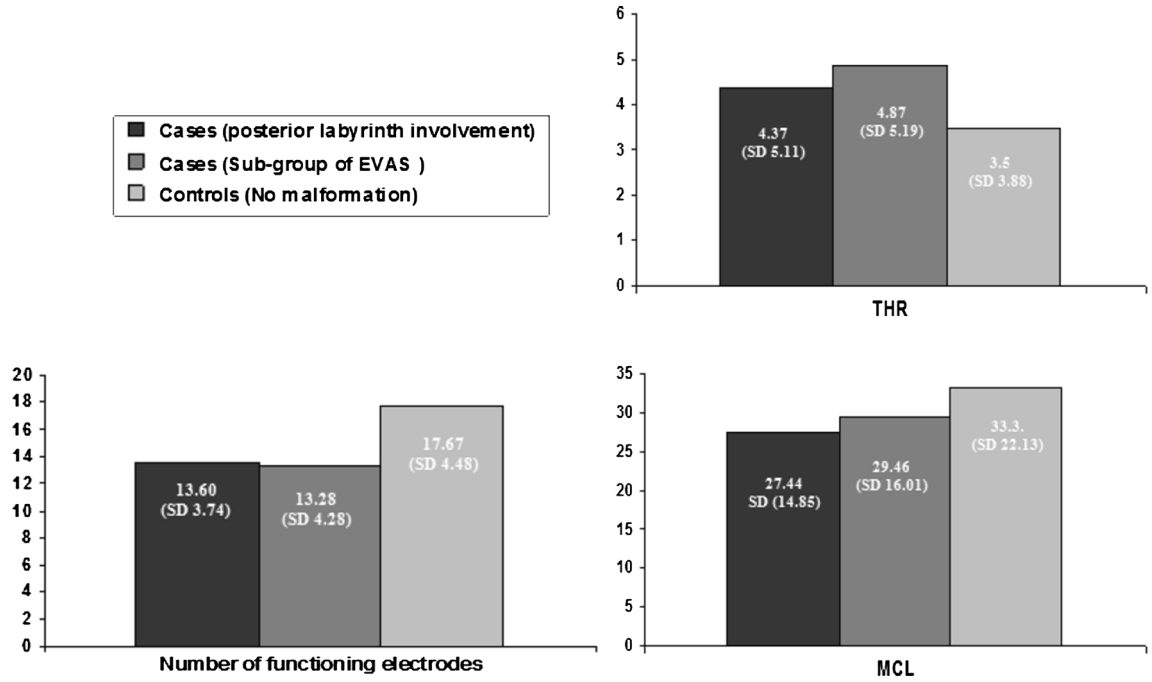


**Fig. 1** Distribution of stimulation thresholds (THR and MCL in microcoulombs) and number of functioning electrodes in the different groups

**Table 3** Distribution of auditory functionality test scores between study groups

| Auditory perception tests at 6, 12, and 24 months[a] | Posterior labyrinth malformations (mean/SD) | No malformations (mean/SD) | Statistical significance ($p < 0.05$)[b] |
|---|---|---|---|
| LIP at 6 | 46.4 % (39.77) | 65.57 % (29.93) | – |
| LIP at 12 | 60.9 % (38.88) | 83.14 % (20.96) | – |
| LIP at 24 | 88.5 % (20.25) | 96.54 % (9.2) | – |
| MTP 3 at 6 | 42.5 % (43.80) | 40.46 % (44.78) | – |
| MTP 3 at 12 | 67 % (40.83) | 81.61 % (27.78) | – |
| MTP 3 at 24 | 79.5 % (33.03) | 98.96 % (4.08) | – |
| MTP 6 at 6 | 20.3 % (34.72) | 25.14 % (35.43) | $p < 0.05$ |
| MTP 6 at 12 | 40.9 % (31.81) | 71.64 % (22.43) | $p < 0.05$ |
| MTP 6 at 24 | 62.5 % (24.02) | 95.29 % (9.98) | $p < 0.05$ |
| MTP 12 at 6 | 1.5 % (4.7) | 13.64 % (27.04) | – |
| MTP 12 at 12 | 27.90 % (40.20) | 40.39 % (35.67) | – |
| MTP 12 at 24 | 37.50 % (38.68) | 86.18 % (18.72) | $p < 0.05$ |
| CLD at 6 | 0 % (0) | 0 % (0) | – |
| CLD at 12 | 20.40 % (33.11) | 17.14 % (30.83) | – |
| CLD at 24 | 42.80 % (45.09) | 75.36 % (25.48) | – |
| OLD at 6 | 0 % (0) | 0 % (0) | – |
| OLD at 12 | 12.50 % (21.24) | 60.07 % (19.87) | $p < 0.05$ |
| OLD at 24 | 32.8 % (32.24) | 63.57 % (30.76) | – |

[a] Subtests were used to evaluate auditory functionality and language acquisition at 6, 12, and 24 months post-implantation

[b] Significant differences ($p < 0.05$), between patients with posterior labyrinth malformations and patients without malformations

with semicircular canal alterations. Various authors, including Valvassory and Clemis [3], Pyle, Romo et al. [4] have reported and studied these malformations. However, although they were first reported more than three decades ago, questions remain about their radiological diagnosis, the factors underlying hearing loss, and their possible association with genetic disorders [18]. These types of malformations associated with profound hearing loss have

good results with cochlear implantation, obtaining a development language similar to patients without these malformations [10].

Bamiou et al., in a CT study, detected EVAS in 60 % of a sample of 116 children with congenital hearing impairment, with semicircular canal disorders (absence or dysplasia) being the second most frequent finding [19, 20]. Other authors report EVAS in 32 % of such cases [9, 21], finding that bilateral involvement was twice as frequent as unilateral involvement [5, 22]. EVAS is considered a characteristic finding in patients with Pendred syndrome [8], which was diagnosed in one of the five children with EVAS in our sample. This malformation has also been reported in patients with nonsyndromic sensorineural hearing loss associated with mutations of the PDS gene (also responsible for Pendred syndrome) [23], and in other syndromes, e.g., coloboma, heart defects, choanal atresia, growth retardation, genital hypoplasia, ear anomalies (CHARGE), Alagille, brachio-oto-renal, and in other inner ear malformations, such as semicircular canal alterations, cochlear dysplasia, or Mondini malformation [9].

The therapeutic approach to malformations with posterior labyrinth involvement, including EVAS, is determined by the degree of hearing loss, and cochlear implantation is indicated in profound hearing loss [24]. It is important to take account of the risk of a perilymphatic gusher during cochleostomy [25], which was observed in two (20 %) of our patients with EVAS but not in any other patients.

Post-implantation data were gathered on the electrical stimulation THR and MCL (in microcoulombs) and on the number of functioning electrodes. There were no significant differences in THR values among groups (4.37 in all cases, 4.87 in EVAS cases, and 3.5 in controls), similar to reports by other authors [5, 7, 9, 14, 26, 27]. No significant differences were found between the patients with malformations and controls in the number of functioning electrodes or MCL, as previous reported [15, 28]. This is because the anterior labyrinth is not affected by these malformations, allowing correct cochlear development and favoring preservation of the tonotopy and cochlear nervous structures. In contrast, higher stimulation THR values are observed in common cavity deformities and cochlear hypoplasia which involve the anterior labyrinth, and worse THR results are obtained after cochlear implantation [28]. In the case of patients with EVAS, however, hearing loss appears to be influenced by reflux from the (abnormally enlarged) endolymphatic sac towards the anterior labyrinth, among other alterations [3, 29]. The endolymphatic sac has immunological and endolymphatic hemostatic functions [30], but it can reflux non-degraded toxic substances and metabolites towards the cochlea, which may explain hearing loss fluctuations. Sudden post-traumatic hearing loss in patients with EVAS may be attributable to the direct transmission of intracranial pressure through the enlarged vestibular conduct towards the inner ear [29]. Other explanations of hearing loss in EVAS patients include damage to ciliated cells from an increase in endolymphatic liquid pressure, associated membranous cochlear dysplasia, or sensory cell dysfunction due to genetic mutations [9, 30]. The role of a conductive component in hearing loss is less clear, although it may be related to stapes hypomobility due to increased endo- and peri-lymphatic pressure, and involving semicircular canal dehiscence, promontory fistula or dehiscent carotid artery, etc. [9].

Various authors have found similar auditory functionality results between patients with and without posterior labyrinth malformations [26, 27, 31–33]. Van Wermenskerken reported auditory perception scores of 48.85 % in patients with posterior malformations versus 54.5 % in those without [33]. The differences between cases and controls were greater in the present study, although the performance by the controls was only significantly better at 12 months in the MTP 6-word and 12-word tests and the OLD test, in agreement with some previous studies [10, 14, 24, 34].

No significant differences in language acquisition were found between the subgroup of EVAS patients and the controls, indicating that they benefited from the cochlear implantation. These patients frequently achieve good performance developing open-set speech perception skills (>24 month use), over 50 % of patients using an exclusively oral communication mode [11, 35, 36].

Anatomical alterations in semicircular canal can produce sensorineural hearing loss, as in the three patients in our sample, and can be associated with other malformations, such as EVAS or cochlear disorders [12, 35–37]. It is therefore of vital importance to use CT to determine whether this anomaly is associated with other malformations in candidates for cochlear implantation [12, 35, 38–40].

The main study limitation is the small sample size, attributable to the low prevalence of this type of malformation, with only 10 cases at our center over the past 5 years. There is a need to verify the present findings in studies with wider samples.

## Conclusion

No significant difference in electric stimulation thresholds was found between cochlear implant patients with posterior labyrinth bone malformations, including EVAS, and those without inner ear malformations. Although some differences in auditory functionality were observed between these groups, cochlear implantation appears to have benefited all of the patients.

Children with EVA malformations have an excellent prognosis for developing open-set speech perception and using oral communication modes.

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