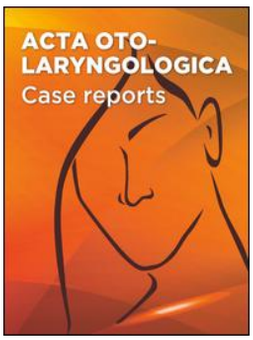

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CASE REPORT

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Pneumolabyrinth following cochlear implantation resolved after shunt adjustment

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ABSTRACT

We present a case of a male with a history of repeated surgeries for a cerebellar astrocytoma, leading to profound deafness and facial paresis on the left side. A ventriculoperitoneal (VP) shunt was inserted to manage hydrocephalus. At the age of 46 the hearing suddenly disappeared on the right side, where he received a cochlear implant (CI). At implant activation, impedances showed atypical high values. One month later impedance levels had further increased and the patient had no benefit from the CI. A computed tomography (CT) scan suggested air in the cochlea (pneumolabyrinth). The shunt was adjusted to elevate the cerebrospinal fluid (CSF) pressure and the pressure in the perilymph to prevent air from entering the cochlea *via* the round window. One year after activation the electrode impedances were normal and the hearing outcome from the implant was successful.

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KEYWORDS
Cochlear implant; pneumolabyrinth; shunt; impedance change

Introduction

A cochlear implant (CI) is a prosthesis for hearing used to bypass a non-functioning inner ear. The first CI was implanted in the 60 s but is today considered an effective routine treatment for patients with severe to profound sensorineural hearing loss. Most patients benefit from the implant and improves in speech perception. However, there are cases when the hearing outcome is not as good as expected and when the rehabilitation process is not straight forward. This can be due to several factors such as long-term deafness, age at implantation or anatomical conditions.

This paper describes a CI recipient with a ventriculoperitoneal (VP) shunt who experienced decreasing benefit after implant fitting. Impedances were postoperative unexpectedly high and varying. The role of the VP shunt in relation to intracochlear air is discussed. In this case more than one factor may have interacted to cause the problems. The implant integrity was tested by Med-El (MED-EL©, Innsbruck, Austria) one month after implant fitting, and it was established that the impedance changes were not due to an implant failure.

This study was approved by the Uppsala Ethical Review Board (Dnr 2019-04203) and informed written consent was given by the patient.

Case report

The patient was a 46-year-old male with a previous history of cerebellar astrocytoma. He underwent initial surgery of the tumour at the age of 3, following which he developed facial paresis and profound deafness on the left side. A ventriculoperitoneal (VP) shunt with a Codman Hakim valve was inserted at the age of 13 to manage hydrocephalus. At the age of 14 a craniocervical fusion was performed to alleviate pressure on the brainstem from a progressive deformity in the craniocervical junction. During hospitalization, due to a severe urinary tract infection with bacteraemia, the shunt was removed. After treatment of the infection no clinical signs of hydrocephalus were present, and reinsertion of the shunt was not deemed necessary. At the age of 37 the high frequency hearing in his right ear started to decline and he received a hearing aid. He also started to suffer from vertigo. A few months later he needed

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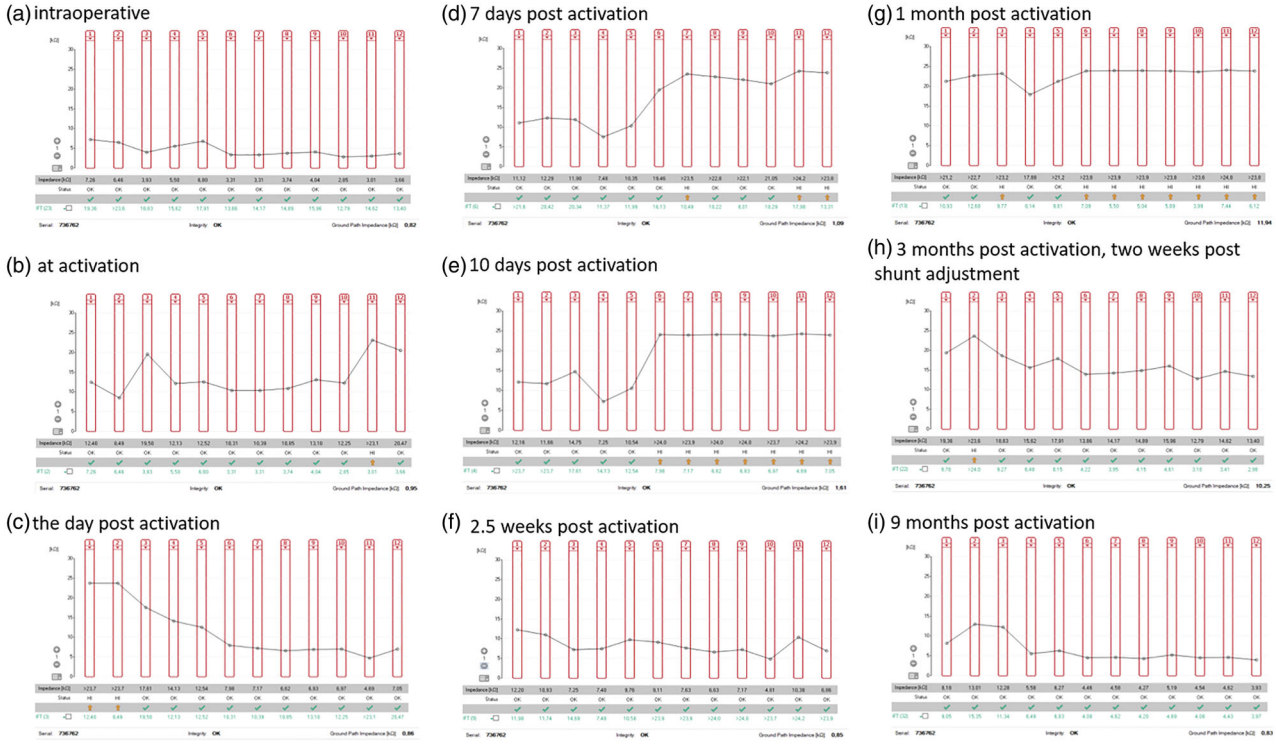

Figure 1. Electrode impedances measured using the Med-El fitting software Maestro (MED-EL©, Innsbruck, Austria). (a) intraoperative – normal values, (b) at activation – electrode 11 HI, (c) the day post activation – electrode 1 and 2 HI, (d) 7 days post activation – electrode 7, 11 and 12 HI, (e) 10 days post activation – electrode 7 to 12 HI, (f) 2.5 weeks post activation – normal values, (g) 1 month post activation – electrode 3 and 6–12 HI, GPI 11.94 kΩ, (h) 3 months post activation, two weeks post shunt adjustment – GPI 10.25 kΩ, (i) 9 months post activation – normal values.

neurosurgical attention again due to a pressure on the brainstem and a transoral dens resection was done. After surgery a cerebrospinal fluid (CSF) leak developed and led to long-term intensive care, and a new VP shunt (Codman Hakim valve) was eventually inserted to manage the leak, deemed in part to be associated with a reactivated hydrocephalus. In the following year the hearing in his right ear deteriorated and at the age of 44 it suddenly completely disappeared within an hour. The high frequency hearing recovered to some extent but was unusable for the patient. The patient now considered himself as bilaterally profoundly deaf.

He was implanted in his right ear with a MED-EL CI (Synchrony FlexSoft 31 mm with 12 electrode contacts) *via* the round window in august 2018. The surgery was performed with no complications and electrophysiology measurements (impedance check, electrical stapedius reflexes eSRT, electrical auditory brainstem responses eABR and auditory response telemetry ART) during implantation showed normal results. The bone bed for the implant was drilled using diamond drills. The depth of the bed was approximately 3 mm. The medial part of the cortex bone towards the dura was kept intact. The whole round window membrane was exposed after the bony overhang of the niche was drilled away. An opening into scala tympani was made using a small needle. The electrode array was thereafter smoothly introduced into the scala tympani. The opening between the electrode and round window membrane was sealed with dried fascia carefully folded around the electrode and glued with tissue glue. The oval window including the stapes footplate were kept intact during the whole procedure. Impedance check from surgery is displayed in Figure 1(a). After surgery, during the healing process, the patient had an infection in the wound which needed two antibiotic treatments.

At the first fitting of the CI system, an impedance check showed a high impedance (HI) value on electrode 11 but otherwise normal values (Figure 1(b)). The patient's subjective hearing was quite well from the start with the implant. The day after, the impedance check showed HI on electrode 1 and 2 (Figure 1(c)) but normal values on the other electrodes. The subjective hearing was still good *via* the implant. Seven days post activation electrode 7, 11 and 12 showed HI, still no deterioration in subjective hearing performance (Figure 1(d)). Ten days post activation electrode 7 to 12 showed HI (Figure 1(e)) and the

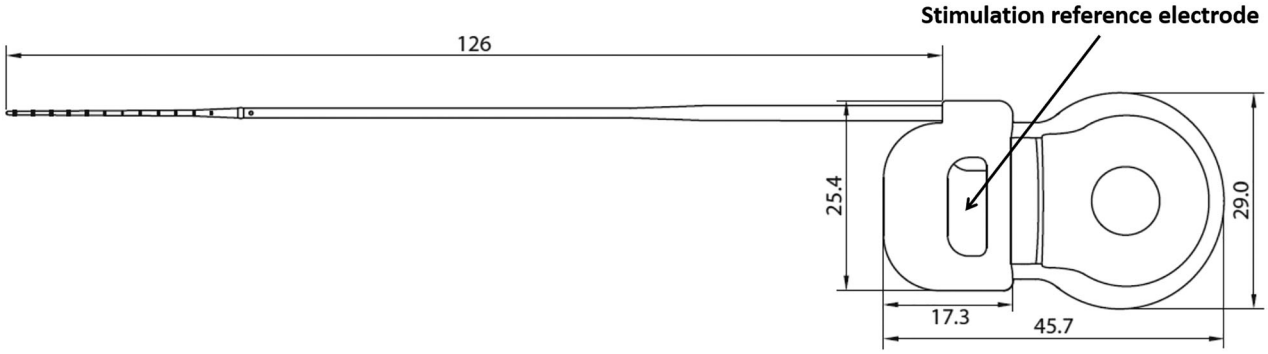

Figure 2. Electrode sketch, stimulation reference electrode (arrow) on the implant housing (MED-EL©, Innsbruck, Austria). Electrode contact 1 is at the tip of the implant array and electrode contact 12 is closest to the implant housing.

patient experienced that the hearing was not as good as it was the first weeks after activation. Two and a half weeks post activation the patient experienced huge fluctuations in hearing performance, the hearing seemed to be better in the morning but worsened during the day. The sound got blurry and weakened in volume. Electrode impedances showed normal values (Figure 1(f)). The hearing *via* the implant, measured in the morning, was at this point 54% on monosyllabic words (MS-words) in free field at 65 dB SPL. The pattern with fluctuating hearing *via* the implant worsened over the following weeks and the patient reported the sound to be as listening through cellophane. One-month post activation eight of 12 electrode contacts showed a HI value and the reference electrode (Ground Path Impedance - GPI), showed a value of 11.94 kΩ (Figure 1(g)). The patient was not able to hear *via* the implant and adjusting MCL: s (most comfortable levels) did not overcome the problem. The GPI value was possible to lower by placing pressure over the CI (over the stimulation reference electrode, Figure 2) but the value went up as soon as the pressure was released. The cause for this behaviour was supposed to be air over the reference electrode, since it is a known and most common explanation for high GPI values. The patient wore a tight bandage over the CI for a week to get rid of the air over the reference electrode but the skin over the implant started to get red and sore due to the pressure from the bandage, and he had to stop wearing it. The patient reported better subjective hearing when lying down, and it remained reasonably good for 30 to 60 min after getting up. The CI hearing seemed to be affected by intracranial pressure as the hearing worsened when standing up, i.e. when the intracranial pressure dropped. A CT-scan showed signs of air in the cochlea around the electrode contacts (Figure 3). No vertigo was described by the patient.

Two possible contributing factors were considered regarding the non-functioning hearing from the implant. The electrode contacts in the cochlea seemed to be surrounded by air instead of perilymph. The air prevented the stimulation from the implant to be transmitted to the cochlear nerve. Air also seemed to be present at the implant reference electrode placed on the implant housing (Figure 2). Air in these two locations isolates the implant electrodes from the inner ear structures they are supposed to stimulate and yields high impedances. This mechanism probably explained the HI values since device failure was ruled out. One possible explanation for air entering the cochlea was the transmission of low intracranial pressure to the perilymphatic system, causing low pressure relative to the middle ear pressure, thereby enticing air to enter the cochlear *via* the round window.

As the hearing seemed to be affected by intracranial pressure fluctuations (i.e. standing up vs lying down), a possible solution was to adjust the shunt to a higher CSF pressure. It was from the start of this episode set to a value of 80 mm H2O and was adjusted up to 100 mm H2O. The adjustment was performed with a Codman Hakim Programmer and verified by the programmer. Unfortunately, the patient the day after the adjustment got pyelonephritis and sepsis, unrelated to the shunt adjustment, which initially prevented him from evaluate what effect the increased shunt pressure had on the implant hearing as he needed hospital care. Two weeks after the shunt adjustment the hearing *via* the implant started to stabilise but he still experienced episodes when the sound completely disappeared, and the sound quality was not satisfactory. An impedance check showed HI on electrode 2 but otherwise normal values. However, the GPI showed a value of 10.25 kΩ (Figure 1(h)). After applying pressure by hand for 3–4 min on the

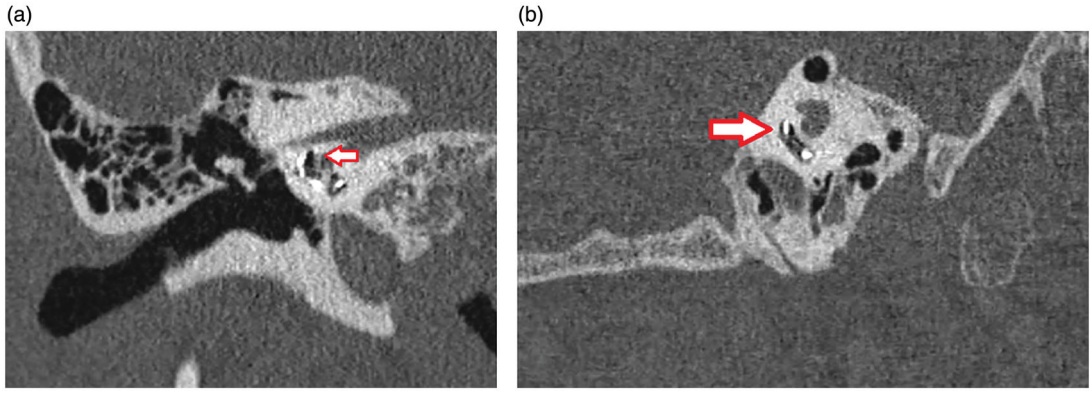

Figure 3. A computerized tomography (CT) scan of the right temporal bone. Note electrode with suspected air (black) surrounding the electrode array in the cochlea (arrow). The suspected air is visible on axial, sagittal and coronal slices, which makes it unlikely that the black signal is an artefact from the metal in the electrode. (a) Coronal view. (b) Sagittal view.

reference contact on the implant housing to get rid of the air we were able to lower that value to 0.85 kΩ which was considered normal. With a GPI under 1 kΩ the hearing *via* the implant was back even though the subjective sound quality was not as good as it was the first weeks after activation. Over the following months the hearing improved and electrode impedances stabilised. From time to time air covered the reference electrode and prevented him from hearing *via* the implant, but he learned how to put pressure on it himself to push the air away. Nine months post activation all the electrode impedances were down to normal values and the problem with air over the refence electrode seemed to be gone (Figure 1(i)). The sound quality was still improving, and he scored 68% on MS-words in free field at 65 dB SPL. Hearing in noise test (HINT) was measured to S/N ratio +3.5 dB. One-year post activation the implant function was still stable and he had no problems with high impedances or air over the reference electrode. He scored 74% on MS-words in free field at 65 dB SPL.

Discussion

This is an unusual case with complications after CI due to air entering the cochlea, described in the literature as pneumolabyrinth [1], as well as air covering the reference electrode on the implant housing. The patient's history, with deficient regulation of intracranial pressure after removal of a cerebellar astrocytoma and subsequent surgeries, is likely to be the main contributing factor to the implant problems. One similar case is described by Moteki et al. [2].

As the patient reported better hearing when lying down or immediately after getting up in the morning, we started to discuss the possibility to elevate his intracranial pressure by adjusting the shunt, to stabilize the physical environment around the intracochlear implant contacts. The CSF space is connected to perilymph *via* the perilymphatic duct, i.e. CSF pressure is transmitted to perilymph pressure [3]. The explanation considered most likely for pneumolabyrinth in this case is air entering the cochlea from the middle ear *via* the round window [4,5]. The low perilymphatic pressure, explained by the patient using a shunt to manage hydrocephalus, makes it easier for the air to enter the cochlea. It is possible, but not very likely, that the external CI magnet could have altered the shunt settings [6,7]. Careful testing was done prior to implantation, with external CI magnets as well as a VP shunt (Codman Hakim valve) of the same type as the patient was implanted with. The conclusion was that it is extremely unlikely that the CI accidently will alter the settings of the shunt, especially as the shunt was placed contralateral to the CI in this case [8].

Whether the shunt adjustment done in this case had any effect on the outcome in the long run is of course unclear. In the case described by Moteki et al. [2] the electrode impedance stabilized after 8 months without any intervention. One possible solution to prevent air from entering the cochlea that was discussed was to do a revision surgery to better seal around the implant where it enters the cochlea through the round window [5]. Other cases of pneumolabyrinth after CI described in the literature were treated conservatively or with revision surgery [2,4,7]. In our case we choose to adjust the shunt settings to speed up the recovery process and avoid another surgery.

It is a known, but not very common, complication that air temporary can cover the reference electrode on the implant housing, especially if the patient has a cold and/or performs Valsalva manoeuvres. An

unusual feature of this case was the intermittent course with multiple unprovoked episodes. The phenomenon seemed to stabilize with time and had disappeared at 9 months post activation.

No signs indicating a problematic outcome were detected during implantation in this case. Even with knowledge of the patient's full medical history, we did not expect these complications. At present, one year after activation, the patient's hearing *via* the implant is above average at our clinic [9], scoring 74% on MS-words in free field at 65 dB SPL.

Conclusion

This is an unusual case with air around the electrode contacts in the cochlea and over the reference electrode on the implant housing. The air in the cochlea disappeared after VP shunt adjustment, verified by normal electrode impedance measures. The air over the reference electrode was recurring, but the patient learned to push it away when it occurred, and over time the problem declined. At present, one-year post activation, the implant function is stable, and the patient has an above average hearing from the implant. Cochlear implant candidates with similar medical history of intracranial tumours with repeated surgeries and VP shunts, even though rare, should be handled with extra caution by the implant team to detect similar complications and extra care should be taken to properly seal the round window.

Informed consent statement

Informed written consent was given by the patient.

Disclosure statement

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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