Cochlear implants in obliterated or ossified cochleas

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Abstract: We present the most recent cases of CI in obliterated or ossified cochleas, describing the most appropriate surgical technique for the insertion of the compressed or double electrode array, highlighting the importance of the combined method (an endaural approach with atticotomy and posterior tympanotomy) to obtain the best view of the promontory. The advantages of this method are that it respects the posterior wall of the EAC in all cases and that it allows the implant to be inserted easily.

Key words: Cochlear implants. Obliterated cochleas. Ossified. Surgical method.

INTRODUCTION

In the case of the slightly permeable cochlea, the insertion of a conventional electrode array is partially or absolutely impossible and in many cases cochlear implantation is contraindicated for fundamental reasons. Firstly, this is due to the difficulty or near impossibility of introducing the electrode array safely; secondly, in the case of cochlear ossification, it is highly unlikely, from the auditory point of view, that effective electrical stimulation will be attained; finally, histological observations seem to demonstrate the poor survival of the neurons of the spiral ganglion, which would make the insertion of the implant somewhat pointless. This occurs, fundamentally in the case of labyrinthitis ossificans caused by meningitis. Nevertheless, it does not occur in cochleas ossified by other causes and, of course, CI is not contraindicated and total ossification is usually limited to the basal portion of the cochlea, the rest of the canal being occupied by fibrous tissue.

We should remember the now historic observations of Schuknecht, who demonstrated, morphologically and functionally, the etiology of sclerosis and labyrinth ossification in the course of chronic otitis media. These observations were subsequently repeated by Charachon, Friedmann and Leroux-Robert, whose histological preparations demonstrate the frequent cochlear affection in these processes, producing sclerosis and ossification. We add that, in the first era of cochlear implants, when Chouard et al implanted their Chorimac with independent electrodes, which were inserted, one by one, through perforations in the promontory, they described the eventuality of finding an obliterated cochlear canal, affirming that this finding did not contraindicate the implant; as Chouard said, it’s better to hear something than nothing.

Various types of electrodes have been designed with a view to exploiting as far as possible the little cochlear capacity offered in this situation. Some designs shortened the electrode array, reducing the free space between the active plates, and others divided it, that is to say, doubling it or splitting it. In some cases, the cochlear problem is due to the continuance of an earlier implant; we have been able to verify this in some of our re-implantations, caused by defects or deterioration of the system over time. The electrolytic phenomena occasioned by the electrical stimulation produces a deposit of calcium and the development of a connective net that makes the insertion of the new implant difficult. The objective of this peculiar implantation is to exploit the existing cochlea to its full advantage, although the prognosis is uncertain and unpredictable with respect to its results.

INDICATIONS AND DIAGNOSIS

On the one hand the antecedents and duration of the deafness can lead us to suspect the existence of a 100% non-permeable cochlea, above all if the patient has suffered from meningitis or if we are dealing with an old, cochlear-type otosclerosis. The CT will confirm the suspicion, although it should be taken into account that this exploration is not conclusive and the help of the MRI is needed. Once the malformation (almost always at the level of the basal turn) and the extent of the cochlear obliteration have been confirmed, we will go on to choose which type of electrode array to use: the compressed or the double array.

There are authors who recommend using the promontory test to evaluate the state of the nervous tract, leading to a contraindication of implantation in
the case of a negative response. We do not carry out this test due to the common possibility of finding “false positives”. The patient may report a hearing sensation, confusing it with the electrical discharge. There was a time when the promontory test was considered to be compulsory for all possible candidates of a cochlear implant. In the case of obliterated cochleas we should consider the usefulness of the promontory test, but not contraindicate the implant if the result is negative. We can only, in our opinion, consider it as relatively indicative with respect to the continuance and possible integrity of the spiral ganglion. The validity of the promontory test, in these cases, should be reviewed. Nevertheless, there is an interesting study by Chouard et al in which they demonstrate the strong electrical resistance in these cases with respect to normal cochleas, drawing attention to the enormous subjectivity of their method and stimulating the round window, looking to avoid the osseous layer of the promontory and to better evaluate the response.

We believe that the best way of evaluating the effectiveness of the promontory test is to accompany it with a functional MRI. By means of this procedure, the test is no longer subjective (sensation of hearing), but becomes, supposedly, “objective and more reliable”, as is proposed by Neumann and Nishida in the updated report edited by Kubo for the Asian Congress on Cochlear Implants. Obviously, according to what emerges from the publications relating to this method, we will be able to make a neural tracing of the acoustic tract and rule out negatives. The use of a positron emission tomography (PET scan) might also be valid. When these methodologies are within the reach of all, many aspects of this controversy will undoubtedly be cleared up.

We consider that the current implantation techniques in the case of ossified cochleas allow for the insertion of the electrode array, in general lines, in the totality of cases and the auditory benefit is always greater than leaving the patient without an implant. We add that the present design of the electrode array facilitates the development of the technique and makes the insertion and use of all the channels possible.

The view of the cochlear obstruction will depend on the radiological explorations and surgical confirmation. Of the first, the most important test is the MRI, better in 3-D, that will allow us to identify three eventualities, which have been very well defined by Chouard. The first, a greater density of the cochlear canal; secondly, images with beaded or fragmented irregularities, and; finally, the absence of liquids in which case the results of the MRI imply total ossification. It case should be reiterated that the CT can provide images of apparent permeability.

Surgical Technique

We use the Lenhardt or the Hannover incision and the means of access to the cochlea does not differ, in these cases, from the conventional, although in the case of implantation with the Nucleus system or the Advanced Bionics 90K, due to the characteristics of these devices, the minimal retro-auricular incision can be made, with utmost ease. Draw attention to the posterior tympanotomy, which must be wide, and pare down as much as possible the posterior wall of the canal, with the objective of obtaining the maximum visibility possible. On occasion, the excision of the incus and of the malleus head associated with the sectioning of the tendon from the malleus to the stapes could be indicated if it is necessary to work on the promontory in its upper portion. The elimination of the posterior wall of the external auditory canal has even been described.

The greatest surgical difficulty lies in the identification of the area of the round window and we have to imagine where it would be found in normal conditions, as the niche of the window is often not recognizable; in these cases, the fundamental point of reference is the oval window and we will make the cochleostomy at some two mm from its lower edge.

Balkany has classified cochlear ossification in three degrees of obliteration: 1. Limited to the niche of the round window; 2. in the basal turn only, and 3. when the obliteration goes beyond this and reaches the middle turn. In the first two cases, the cochlear lumen is found after the drilling of the promontory, in an ascendant direction, always bearing in mind its anatomical situation. It can often be seen that the material that occupies the lumen is of a different composition to that of the otic capsule; it is softer and clearer in color and, on occasion, it can be extracted. On other occasions, this tissue is compact and, therefore, it is impossible to view the cochlear lumen clearly and it is necessary to drill in an anteromedial direction until finding the lumen of the useful cochlear space. Nevertheless, complete ossification is less frequent; in their series of 24 cases, Green et al only found it in two. Furthermore, in their series, Cohen et al were obliged to carry out a partial insertion of the implant in 7% of the cases (out of 110) varying the number of active electrodes between 10 and 18 as they were dealing with semi-obiterated cochleas with radiological images of normal appearance.

When the ossification goes beyond the basal turn and reaches the middle turn, more aggressive techniques have been described. Both Gantz and Steenerson have proposed the creation of a perimodiolar canal to hold the electrode array. Subsequently, Telian described a similar procedure,
making use of a wide mastoidectomy and elimination of the posterior wall of the canal to achieve a clear view, the widest possible, of the promontory. The obliteration of the cavity is achieved with an aponeurotic muscle flap or by sacrificing the membranous EAC as we do when we operate on tumors of the middle ear.

The best manner, without eliminating the posterior wall of the canal, is to carry out a mixed or combined approach. (Figure 1). This combined method includes a posterior tympanotomy and, from the front, an endaural approach, as proposed by Dr. Arauz of Buenos Aires, based on the technique described by Banfai and al for the insertion of their primitive 16-channel extracochlear implant. Remember that this author devised and promoted a device in the shape of a plate that was placed directly on the promontory, as Banfai said, to avoid endocochlear damage as a consequence of implantation. This implant was not widely used but we are indebted to Banfai for the description of the technique and a magnificent study of cochlear anatomy in which its reference points are defined.

Once the mastoidectomy and the posterior tympanotomy have been carried out, the area of the round window is accessed; if it is not possible to identify the round window, the drilling will be carried out at some 2 mm from the oval, in an anterior and slightly ascendant direction, keeping in mind the proximity of the carotid canal and the anatomical inclination of the cochlea. Whatever the ossification or partial obliteration should be, after the study of the data provided by the CT and MRI, we must choose the electrode array we will use; the reduced or shortest (compressed), or the split or double array guide. This was used in one of our cases.

If we opt for the split electrode array it is almost always feasible to insert both its components through the posterior tympanotomy. We will perform the cochleostomy in the basal turn on the promontory at some 2 mm above the area of the round window. The cochleostomy in the middle turn will be made in front of the oval window.

If we opt for the promontory implantation with a compressed electrode it is necessary to widen the access in an anterior direction, peeling away the membranous EAC and lifting, in a forward direction, the tympanic membrane flap, as is done in the tympanoplasty. This maneuver will give us ample visibility of the promontory, but we must widen it by sectioning the malleus head, the tendon of its muscle and eliminate the incus, to obtain a more comfortable surgical field.

Once the anterior view has been attained, the drilling made previously can be seen -through the posterior tympanotomy- of the area of the round window and, just above and in an ascendant direction, we will cut a niche following a transversal line almost parallel to the tube in its third portion, until reaching the level of the stapes. We will then pass on to drill in the opposite direction and, finally, ascending towards the cochlear apex, we will have three lines cut in the shape of a “Z”. While the formation of this cochlear neocanal is being carried out, it is very important to leave some osseous bridges that will impede the subsequent displacement of the electrode array. The other method is to cut the neocanal in the shape of a “C”. To do this, from the area of the round window, once the obliterated cochlear canal has been localized (almost always full of fibrous tissue which can easily be resected with the stilette) we will follow upwards until we are 2 mm in front of the spoon-shaped apophysis, to descend and arrive at 1 mm from the inferior edge of the oval window.

We must warn that, should the cochlea be totally ossified, it is advisable to make the cut in the shape of a “Z” as there are no visible cochlear references. The osseous bed for the receptor will then be made in the mastoids and will be fixed in the conventional manner. Holding the receptor in place will facilitate the surgical maneuvers required for the insertion of the electrode array.

We should caution that partial insertions of conventional electrode arrays should be avoided and we would advise that an implant devised for the particular purpose should be obtained, with a view to achieving maximum effectiveness. The initial insertion of the device will be carried out from behind and

![Figure 1. Combined approach: transmastoid and transean](image-url)
through the posterior tympanotomy; we will then change our approach to work from the front directly on the promontory and introduce the electrode array in the canal we cut earlier and beneath the osseous bridges made previously. The manipulation of the system must be very careful in order not to damage any microplate. Finally, whichever device is used, it will be covered with a fascia lamina secured with fibrin glue (Tissucol®). The use of glue must be avoided as these will damage the electrodes.

OUR CASES

Case 1:
A 72-year-old woman diagnosed with cochlear otosclerosis. The disease began when she was 20 and its evolution sped up after her second pregnancy, obliging her to use a hearing aid from then on as a stapedectomy was not indicated due to the sensorial affectation of the disease. She came to our Cochlear Implantation Center because her hearing aid did not help her in communication. Furthermore, she suffered from bilateral tinnitus.

The CT and the MRI showed the existence in both cochleas of obliterated or ossified tissue, giving a beaded cochlear image in the resonance image (Figure 2).

It was decided to insert a Combi 40+ GB double cochlear implant in the right ear, with two electrode array guides (Figure 3): that of 7 mm in the basal turn and that of 5 mm in the middle turn. The surgery was carried out without major difficulties, with a postoperative evolution similar to that of the majority of implanted patients, the patient being discharged after 48 hours.

In the telemetric revision all the electrodes had good impedance, but the patient only obtained benefit from the basal electrodes.

Case 2:
A 58-year-old male with right progressive profound hearing loss since infancy, caused by recurrent suppurative otitis media.

He suffered from pneumococcal meningitis in February, 1994, with sequelae of left-side hearing loss. In 1995 a right-side cochlear implantation with a Digisonic device was attempted; this was unsuccessful due to the patient’s having fibrosis in the basal turn. A second operation was carried out 14 days later, as the MRI showed partial but sufficient cochlear permeability, for which reason the cochleostomy was widened in an ascendant and forward direction, and a partial insertion was achieved.

Over the course of time, progressive deterioration occurred in the CI, it was decided that the implant should be replaced but this could not be done because the cochlea was obliterated, without useful promontory

Figure 2. CT and MRI: Partial beaded cochlear obliteration

Figure 3. Implant with two Med-El GB 40+ electrode array guides. Double cochleostomy on basal and middle turns.

Figure 4. Complete bilateral cochlear obliteration: post-otitis in the right ear and post-meningo labyrinthitis in the left ear.
surgical references. The MRI showed complete obliteration (Figure 4). Given the circumstances we decided for safety reasons to operate on the left ear, with an obliterated cochlea, but with surgical references intact.

After the sectioning of the malleus head, the incudostapedial disarticulation and the extraction of the incus, a letter “Z” was drilled in the cochlea and a Combi 40+S implant inserted through it (Figure 5).

This intervention is much more laborious than that of the double cochleostomy. It is essential to take special care to respect the carotid artery in the antero-inferior portion of the promontory and the tympanic portion of the facial nerve, in the upper part. Nevertheless the post-operative evolution did not present any differences with respect to other implants.

Case 3:

A three-year-old boy who was referred to us after the failure of an attempted cochlear implantation for profound bilateral hearing loss, after suffering from pneumococcal meningitis a year and a half earlier, complicated by subdural left-side parietooccipital hygroma, which required neurosurgical treatment.

The temporal bone CT was repeated, showing apparent normality in the cochlear permeability.

The child was operated on again and fibrosis and complete cochlear ossification could be seen despite the radiological reports, for which reason a “Z” was drilled in the promontory, following a combined approach, and a Nucleus 24 Contour was implanted. A response was obtained with 8 electrodes. The post-operative evolution was very good.

With reference to the functional results, we verified that in the first case, in which a double electrode was inserted, even with acceptable impedances, the first four electrodes were not useful. The subsequent radiological study showed good insertion of this portion of the electrode array; it had not been pushed out as might have been expected.

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In the second case the result was poor: the tests show thresholds at 30 dB, but this only serves for the detection

C40+ (24 electrodes across 27.4 mm)

C40S (24 electrodes across 13.1 mm)

C40+ GB (14 electrodes across 6.6 mm)  
(10 electrodes across 4.4 mm)
of sources of sound and for a slight improvement in the recognition of phrases with the help of lip-reading.

In the third case the result has been surprisingly good: the tests show thresholds at 42 dB; however, the child is advancing notably in acquiring auditory skills and is developing an oral language similar to that of children of the same age with a cochlear implant.

**DISCUSSION**

For the evaluation of the results and effectiveness of the device, we believe it important to separate this group from that of normal implants and establish a universal criteria of evaluation.

The mediocrity of the results obtained in these cases can be considered to be directly linked to the number of nervous fibers, embedded in fibrous-bone tissue, that lead to a deterioration of the spiral ganglion. That is to say, only those that have remained more or less unharmed by the pathology can make good use of the electrode array, the different electrical impedance offered by the bone being another contributing factor. The resistance factor is possibly the most important and in fact the necessary high incidence of stimulation that frequently leads to diaphonic interaction between electrodes and a very tight dynamic range has been shown. This is also facilitated by the greater proximity between the microplates. For this reason the judgment as to the effectiveness of the CI must be very prudent with respect to the evaluation of the results and some are more pessimistic than others, perhaps because they are always compared with cases of normal insertion of the implant. The optimism of Kemink, who in five children with partial insertion of the device affirmed results similar to those of normal cochleas, stands out in contrast to the series of cases described by Balkany and Parisier, which had much worse results. We consider that the group of implanted patients with obliterated cochleas should be considered apart from and different to that with permeable cochleas and we would never dare to predict its prognosis.

**CONCLUSION**

Cochlear ossification does not contraindicate CI; however it does obstruct it and make it more difficult; electrode arrays designed specifically for this purpose should be used to overcome this difficulty. Furthermore, prognosis is uncertain with respect to the discriminative results of the language and in relation to the increase in electrical resistance and the functional state of the spiral ganglion.

With respect to which technique to use, we recommend a mixed method combining an endaural approach with atticotomy and posterior tympanotomy, to achieve easier access to the cochlear area and to better recognize the points of anatomical reference.

It is usually impossible to insert a classical electrode array; it is necessary to use a compressed or double model, avoiding partial insertions of a conventional device (Figure 6).

We add that it is obligatory to carry out an MRI in all cases, above all in those cases with a history of meningitis, as the CT may offer false negatives.

**References**