

Successful partial cochlear implantation in a patient with relapsing polychondritis

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Abstract

The author reports on a case of relapsing polychondritis in which partial insertion of a cochlear implant has been successful. A woman with known relapsing polychondritis gave a history of increasingly severe auditory loss of sensorineural type occurring bilaterally. Previous repeated treatments with corticosteroids and immunosuppressive medications had been attempted. A right-sided cochlear implantation with partial electrode array insertion was carried out due to the patient's cochlea being obliterated. The outcome was excellent and the improvement in

hearing was maintained. Relapsing polychondritis (RP) is a rarely-seen disorder, the aetiology of which is obscure, but which results in inflammation within cartilage throughout the entire body. The cochlea may become obliterated by this process. Cochlear implantation may be needed in such circumstances. The successful outcome in this case shows a potential role for cochlear implantation in individuals with inner ear disorders produced by immune disorders or relapsing polychondritis.

Keywords: Polychondritis, relapsing, sensorineural hearing loss, cochlear implantation.

Jaksch-Wartenhorst may have been the first person to describe the condition currently termed relapsing polychondritis (RP), writing in 1923.^[1] RP is a rare condition which features recurrent exacerbations of inflammation affecting cartilage. The pathogenic basis and aetiology of RP is uncertain. Some 40-50% of individuals with RP develop conditions affecting the inner ear at some point in the disorder.^[2,3] This report is of a case of RP in which severe auditory loss had occurred, affecting both ears, and of sensorineural type. Previous treatment with corticosteroids and immunosuppressive medications had been to no avail. A cochlear device was implanted successfully, with partial electrode placement.

Case report

A woman aged 53 years was referred to an ENT clinic in 2004 with a history of profound auditory loss of sensorineural type occurring bilaterally, secondary to RP. She also had vertigo. Total deafness had been present in the left ear for 15 years, and in the right for two months. A diagnosis of RP was reached due to chondritis of the nose, a non-erosive polyarthropathy with negative serology, abnormal vestibulo-cochlear function and appropriate histopathological appearances. Pure-tone audiometric analysis indicated bilateral total deafness. Neither corticosteroid nor immunosuppressant treatment had restored hearing. As expected,

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there was no response on either auditory brain stem response or distortion product otoacoustic emission testing. The temporal bone had been imaged using computed tomography (CT) in 2004, as shown in Figure 1. The appearances were interpreted as indicating an intact cochlea on the right, whilst also showing that the left cochlea was partially obstructed.

It was proposed that a cochlear device be implanted in the right ear. An aortic aneurysm required surgery before the device could be implanted, entailing a delay until 2006. The temporal bone was re-scanned (Figure 2), the appearances at that point showing partial obstruction of the basal turn of the cochlea on the right side, and a more significant obstruction on the left. Right-sided cochlear implantation was performed in 2006 using a Digisonic SP cochlear implant from the Neurelec Company. As we found an obliterated basal turn in the cochlea, the device was partially introduced starting at the second turn of the cochlea. Seven electrodes out of 20 were introduced, and their placement confirmed by transorbital-ocular view.

One month later, the Digisonic SP processor was fitted. Six electrodes were found to be active. Pure tone audiometry showed good thresholds of around 20 dB between frequencies of 250 - 4000 Hz, with the cochlear implant operating in a free field. Intelligibility was around 30% with the cochlear implant alone. The patient reported a high level of satisfaction with the device, which she was using throughout the day. In 2011, the patient benefited from an upgrade to a Saphyr SP processor from the Neurelec Company. There was a definite improvement in the result obtained. Intelligibility with Lafon lists of monosyllabic words was around 50% at 65dB with the cochlear implant alone, and reached 94 % with the cochlear implant used in conjunction with lip reading. The score on the MMBA battery sentences was 41% at 65dB with the cochlear implant alone, and 90% with the cochlear implant used in conjunction with lip reading. The patient scored 205 on the Nijmegen cochlear implant questionnaire.

Discussion

RP is a disease affecting multiple body systems and has an association with autoimmune disorders such as Sjögren's syndrome, systemic lupus erythematosus and rheumatoid arthritis. The condition was first recognized by Jaksch-Wartenhorst in 1923, being described as a poly-chondropathy.^[1] RP presents with chondritis affecting the pinna, joint inflammation, symptoms affecting the larynx

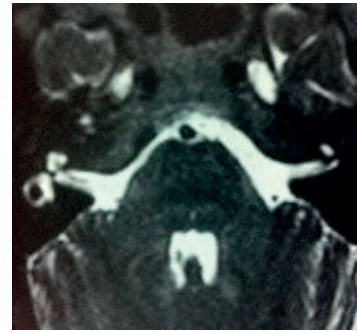


Figure 1. The CT image shows a normal cochlea on the right side and a partial obstruction on the left side.

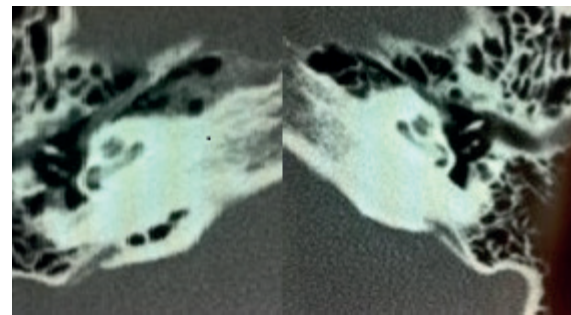


Figure 2. The CT image shows a partial obstruction of the basal turn of cochlea.

and trachea, chondritis of the nose, eye inflammation, symptoms arising from the audiovestibular apparatus and cardiovascular problems.^[4] The rate of inner ear complications in RP sufferers is thought to be 40-50%. Auditory loss of sensorineural type may affect one or both sides and the loss may worsen slowly or very rapidly, with resulting cophosis.^[5,6] Exactly how the inner ear complications unfold is not currently known. Given that the inner ear does not contain cartilaginous tissue, it is theorised that a different mechanism must be responsible, such as vasculitis of the internal auditory artery, affecting the cochlear or vestibular branches.^[2] Hoshino et al^[7] examined temporal bone taken from a case of RP in which deafness had rapidly occurred in both ears. They suggest that viral labyrinthitis occurring alongside RP led to cophosis, as the histological appearances showed encapsulation of the tectorial membrane, and severe degeneration of the organ of Corti, as well as of sensory neurons. Miyazawa et al^[8] used immunohistochemical techniques to highlight type 2 collagen in

histological sections obtained from three cases of RP in which RP had led to sensorineural type auditory loss. They discovered that degeneration particularly affected type 2 collagen in the tectorial membrane, where it is abundant, and thus, they suggest, degeneration can be linked to auditory dysfunction. Type 2 collagen is found in various structures of the external and middle ear: the pinna, the cartilage supporting the external auditory meatus, the ear drum and the annulus of the ear drum.^[9]

Issinget et al ^[10] performed serology for anti-labyrinthine antibodies, finding these were positive in a case of RP where there was impairment of auditory and vestibular function. Circulating immunoglobulins or T lymphocytes targeting the self may produce an inflammatory response and apoptosis in the inner ear. Schuhknecht ^[11] observed that the inner ear lacks cartilaginous structures and suggested that a destructive vasculitis of the labyrinthine artery, or branches thereof, was the pathological mechanism. Such an explanation fits with the observation that systemic necrotizing vasculitis can occur in RP.^[12] Whilst auditory loss of sensorineural type does affect 40-50% of RP sufferers, the majority of such cases do not develop severe auditory impairment.^[13] Usually, the therapeutic interventions used for RP, i.e. nonsteroidal anti-inflammatory drugs, corticosteroid and immunosuppressive agent administration, do partially rectify or protect auditory function in RP cases.^[14] But rare cases also occur in which auditory loss secondary to RP is not responsive to the usual therapeutic measures. Implantation of a cochlear device has been found beneficial previously in cases of auto-immune ear disorders, namely systemic lupus erythematosus, Cogan's syndrome, Buerger's disease and related disorders.^[15] Cochlear implants work by stimulating the para-cochlear spiral ganglion directly. They are potentially highly effective in cases where cochlear function is impaired. One such scenario would be RP resulting in cophosis. Nevertheless, as may happen when there is ossification of the cochlea for other reasons, such as meningitis, cochlear obliteration may render

the implantation of a cochlear device highly challenging. Thus, a high level of clinical vigilance is needed in any case where cochlear obliteration is deemed a risk, so that the device may be implanted prior to the point where cochlear obliteration prevents the siting of electrodes. In the case reported here, good results were still obtainable, despite only partial implementation (five electrodes out of 20 were possible). This outcome is highly encouraging, as the results are superior to other possible options, including an auditory brainstem implant. In clinical practice, whenever possible a cochlear implantation is preferable, even if only partial, to an auditory brainstem implant.

Conclusion

Relapsing polychondritis is a rarely seen disorder, the aetiology of which is obscure, but which results in inflammation within cartilage throughout the entire body. The cochlea may be obliterated by this pathological process. Cochlear implantation may be needed in such circumstances. The successful outcome in this case shows a potential role for cochlear implantation in individuals with inner ear disorders produced by immune disorders or relapsing polychondritis.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

Author Contributions: Designing the study – B.A.; Collecting the data – B.A.; Analyzing the data – B.A.; Writing the manuscript – B.A.; Confirming the accuracy of the data and the analyses – B.A.

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