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Cochlear implantation in a case of Wegener's granulomatosis

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Abstract

Cochlear implantation is now a routine clinical procedure for deaf patients in many countries (Gibson, 1987). It replaces the function of damaged cochlear hair cells and, therefore, detects sound and by electrical stimulation produces an appropriate signal in the remaining cochlear nerve fibres (House and Berliner, 1991). Wegener's granulomatosis is an uncommon auto-immune disease. It has a peak incidence at the fifth decade with slight male predominance. Nasal problems are the predominant presentation with otological manifestations presenting rarely. We report a case of Wegener's granulomatosis presenting with total hearing loss and after right cochlear implantation a free field threshold of 40 dB and 20 per cent Bamford-Kowal-Bench (BKB) speech test.

We conclude that Wegener's granulomatosis is not an absolute contraindication for cochlear implantation.

Key words: Wegener's granulomatosis; Cochlear implant

Introduction

Cochlear implantation has received a good deal of publicity as a method of rehabilitation of deaf patients who do not derive benefit from amplification.

Tests of speech understanding are often required in evaluation of schemes of aural rehabilitation. BKB standard sentence lists were compiled from the utterances of hearing-impaired children (Bench and Bamford, 1979).

They contain straightforward vocabulary and syntax and the standard test comprises 21 lists, each with 50 key words in 16 sentences. Because of the ease with which the lists can be administered, and because there are a large number of them, the lists have been used extensively to measure speech reception skills and benefits from amplification both with children and adults.

Wegener's granulomatosis was first described in 1936 (cited in Komblut, 1982) as a necrotising granulomatous disease affecting the upper and/or lower respiratory tract and associated in classic cases with focal glomerulonephritis. The nasal and paranasal cavities are usually the site of the presenting lesion (Friedmann and Bauer, 1973). Because of this, the otolaryngologist plays an important role in the early diagnosis and treatment (McDonald and DeRemee, 1983). Although its exact cause is unknown, it is assigned to the auto-immune diseases (Illum and Thorling, 1982). As it is a generalized disease, the kidneys, eyes, lungs, joints, skin and the nervous system may be involved. McDonald and DeRemee (1993) reported head and neck involvement in 72.3 per cent of their review of 411 patients with Wegener's granulomatosis. They also reported involvement of lung in 71.2 per cent and of kidney in 57.5 per cent in the same series. Although otological involvement has been reported in the literature, it is uncommon for it to be the presenting problem (Murty, 1990). However, three cases presenting with otitis media with effusion were reported by Karmody (1978). Serous otitis media was the initial manifestation of the disease in seven cases, out of a review of 112 patients with Wegener's granulomatosis (McCaffrey *et al.*, 1980). It was secondary to nasal and eustachian involvement. They also reported that nine patients had significant sensorineural hearing loss. Treatment with prednisone and cyclophosphamide produced improvement in the hearing in five patients. The exact cause of the sensorineural hearing loss in Wegener's granulomatosis is not clear. It may be due to vasculitis of the cochlear vessels, granuloma pressing on the acoustic nerve, and deposition of immune complexes in the cochlea. Otological involvement with Wegener's granulomatosis has been reported by many authors, but they do not describe it either as the initial problem or subsequent to nasal involvement (Fahey *et al.*, 1954; Blatt and Lawrence, 1961; Per-Lee and Parson, 1969; Novack and Pearson, 1971; Linthicum and Schwartzman, 1972; Bourdiniere *et al.*, 1973; Friedmann and Bauer, 1973).

To our knowledge, no one has described cochlear implantation in Wegener's granulomatosis. This is a report of a patient who presented with total hearing loss and was implanted without any surgical or healing problems.

Case report

A 71-year old male presented in May 1990, to our outpatient clinic with history of sudden hearing loss, no pain and no fever. On examination the tympanic membranes were intact, and no abnormalities of nose or throat were detected. An audiogram showed a right sensorineural hearing loss of 30–50 dB (Figure 1), and left mixed hearing loss of 80–90 dB (Figure 2). A hearing-aid was advised. After two months, the patient came back with the same history. Clinically, the left tympanic membrane demonstrated serous otitis media, and a tympanogram showed a type B curve. Left myringotomy was performed the

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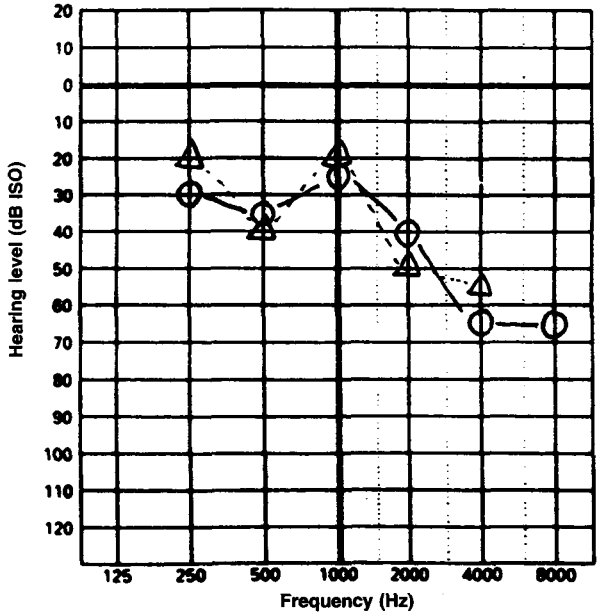

FIG. 1
Right sensorineural hearing loss of 30–50 dB

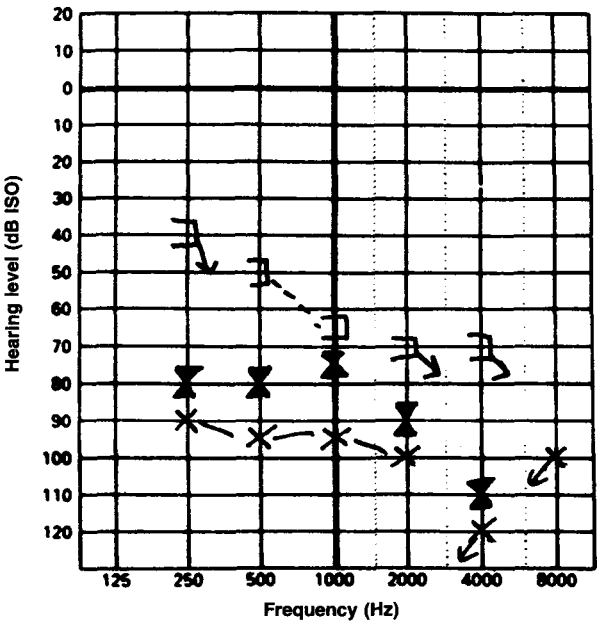

FIG. 2
Left mixed hearing loss of 80–90 dB

following month, straw-coloured fluid was aspirated and a t-tube was inserted, the postnasal space was examined endoscopically and a biopsy was taken blindly revealing the histological picture of chronic non-specific inflammation. All other investigations were normal including chest X-ray, 4 mmol/l serum urea, 57 μmol/l serum creatinine, 70 g/l total protein, 72 per cent granulocytes and 21 per cent lymphocytes. Twenty-two months later he presented again with left otalgia, aural discharge and bilateral sudden total hearing loss. On examination discharge was seen coming out of the inserted t-tube from which a swab was taken. There was no presence of bacterial growth, but an audiogram showed no residual hearing on both sides (Figure 3). Hearing therapy was offered. After four months he presented at the ophthalmological outpatient clinic with right orbital swelling and pain. Wegener's disease was confirmed by a high titre of 1/320 positive anti-neutrophil cytoplasmic antibody (ANCA) and 72 per cent neutrophils. There was no renal or chest involvement. Serum urea was 5.6 mmol/l, serum creatinine was 69 μmol/l, total protein was 70 g/l and the chest X-ray was normal. The disease was controlled with cyclophosphamide 100 mg three times per week and prednisolone 10 mg orally daily. He was referred to the cochlear implant team to assess suitability for implantation.

On evaluation of his suitability for implantation it was considered he was unable to benefit from his hearing aids worn previously. He had been receiving weekly therapy from a hearing therapist to help both his lip reading and overall communication skills. His speech preservation was good. His intonation patterns were good but his pitch range was reduced. He had accepted his deafness and had a realistic expectation of how a cochlear implant could

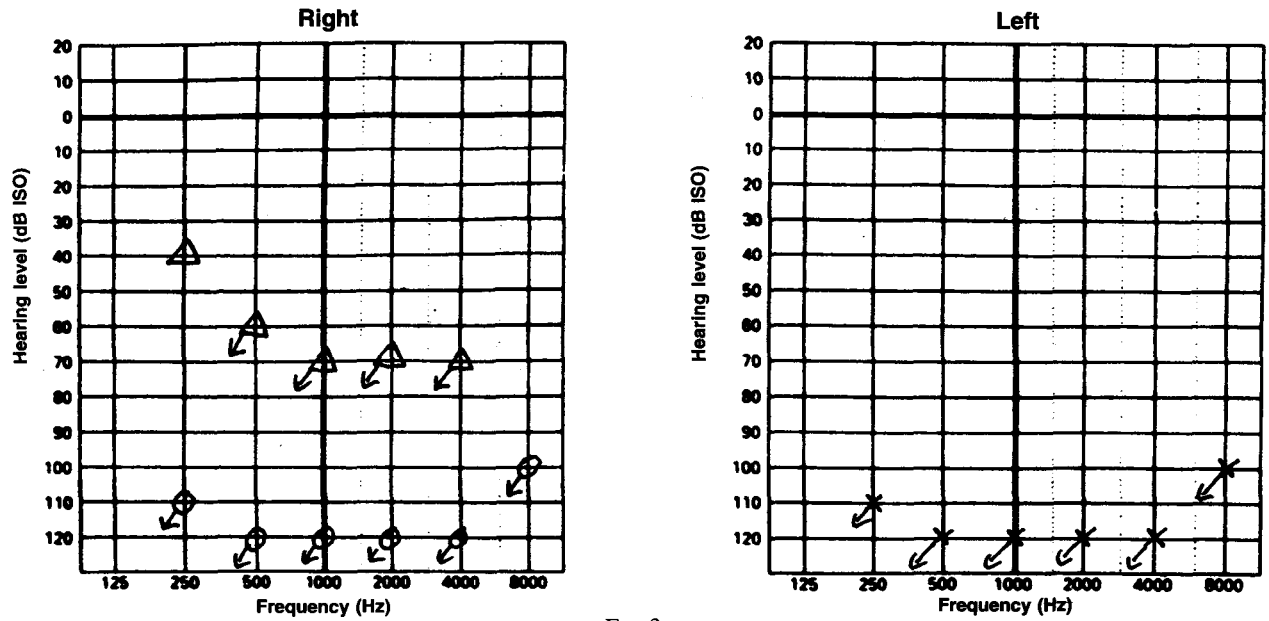

FIG. 3
Bilateral total hearing loss

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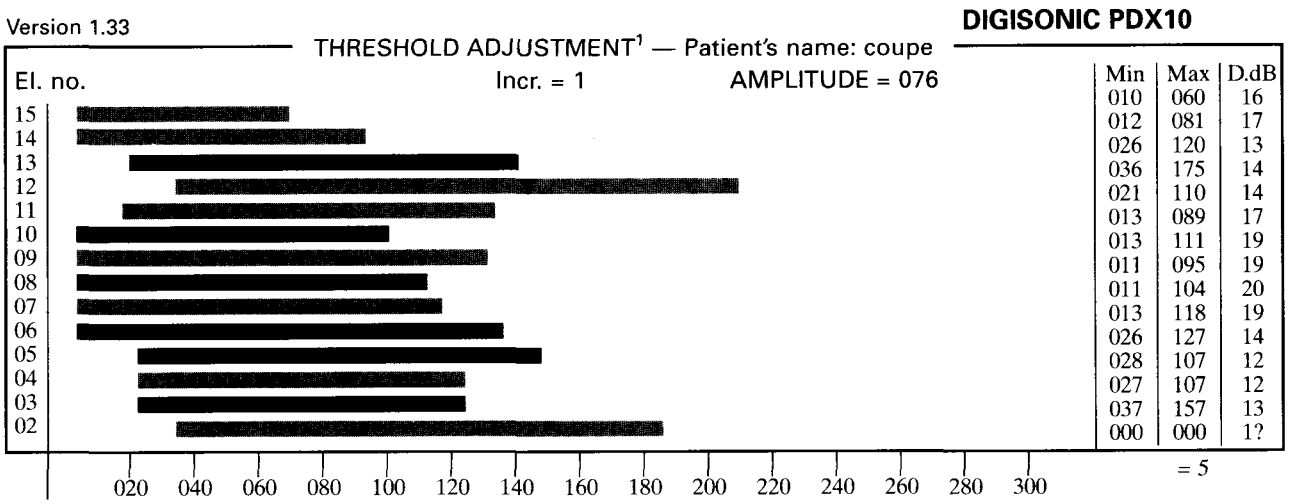

FIG. 4
Dynamic ranges after implantation of Digisonic 15 electrode cochlear device.

improve his quality of life. Computerized tomography (CT) showed both cochleas to be normal.

The following month a Digisonic 15 electrode cochlear device was implanted easily and successfully in the left ear (cortical mastoidectomy and posterior tympanotomy approach). The clips were removed after seven days. The patient was home four days after surgery. He has not developed any surgical complications following his implantation. Fortunately, at his hearing assessment nine months after implantation, he had a free field audiogram of 40 dB hearing loss and 20 per cent BKB speech test in the right implanted ear.

The patient wears the implant for 15 hours each day, he has 14 electrodes active, all with good dynamic ranges (Figure 4). He scored well on the gap detection test, obtaining a score of 65 (71 per cent). This test measures the minimum duration of a silent interval which a patient can detect in the middle of a burst of noise. The duration of each burst is a second. Gaps of 5 ms, 10 ms, 20 ms, 40 ms, 80 ms, 160 ms and 320 ms are used. Each gap is presented 10 times in a random order. On VCV (vowel-consonant-vowel) testing, he correctly identified 22.9 per cent of consonants on lipreading alone. This score increased to 27.1 per cent when the implant was switched on. This test measures the accuracy with which consonants can be identified in vowel-consonant-vowel nonsense words. The test consists of a block of 48 trials in which each consonant occurs four times. On CDT (connected discourse tracking), his score was 59 words/three minutes for lip reading alone and 74 words/three minutes when lip reading was combined with electrical stimulation from the implant. This test measures a patient's ability to understand connected speech. He was able to correctly identify nine out of 20 environmental sounds from a tape. His intonation patterns have improved and his pitch range has increased.

Discussion

Wegener's granulomatosis is an uncommon disease in adults with a peak incidence at fifth and sixth decades of age. It is rare in childhood with a slight male predominance (Nicklasson and Stangeland, 1982). It has a characteristic clinical presentation with nasal and paranasal lesions predominantly but pulmonary and renal involvement is also common. Otological manifestations are almost always secondary to nasal involvement. The incidence of otological involvement varies widely from 19 to 61 per cent (Murty, 1990). McDonald *et al.* (1981) divided otological involvement into three basic types:

(1) Serous media due to Eustachian tube blockage and nasopharyngeal involvement.

(2) Destruction of the middle ear and mastoid cavity by Wegener's granulomatosis.

(3) Sensorineural defect due to inflammation of the cochlear vessels.

Its presentation as aural symptoms is uncommon and to be presented as the only problems is rare. Fauci *et al.* (1983) found only five patients out of 85 in their series complaining of aural symptoms. A case of Wegener's granulomatosis limited to the ear without involvement of the other systems was reported (Ito *et al.*, 1991). In this case, clinical and laboratory and pathological investigations were consistent with Wegener's granulomatosis from the initial presentation.

It is recognised that Wegener's granulomatosis is a fatal condition, unless treated early, and its diagnosis depends on clinical and histopathological features. The recommended investigations include high sedimentation rate, positive ANCA test, proteinuria, abnormal serum creatinine level, anaemia, and nasal biopsy or middle ear mucosal biopsy, if indicated, should be taken for histopathological analysis. Finally, if the diagnosis is still in doubt after these investigations a renal biopsy could be considered. Once diagnosis of Wegener's granulomatosis has been confirmed, cyclophosphamide and steroids should be started. If no improvement in the hearing level of the patient has been recorded after improvement of the disease (negative ANCA test), evaluation for cochlear implantation in cases of total hearing loss should not be delayed.

We conclude that Wegener's granulomatosis is not a contraindication for cochlear implantation.

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