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Cochlear implantation of a Hungarian deaf and blind patient with discharging ears suffering from Behçet's disease

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Abstract
A case is reported in which a Nucleus 22 channel intracochlear device was implanted a deaf/blind Hungarian adult with discharging ears suffering from Behçet's disease. Preconditioning surgery was employed three months prior to the implantation procedure to ensure a sterile, dry protected environment for the electrodes. One month after implantation, the patient exhibited excellent auditory discrimination capability at the time of the first switch on. We suggest that some deaf/blind individuals may serve as very good candidates for intracochlear implantation.

Key words: Cochlear implant; Behçet's syndrome

Introduction

Cochlear implantation in normally-sighted individuals is generally regarded as a medical strategy which serves to supplement lipreading skills. Cochlear implantation of the deaf and blind patient was therefore initially considered an intervention of doubtful benefit and there have only been a limited number of implantations round the world. Nevertheless, when the onset of blindness precedes that of deafness, the adaptive plasticity of the cortex, together with the motivation of the patient may recruit the deaf/blind patient into a unique group of excellent implant performers (Ramsden *et al.*, 1994). This case report suggests that multichannel intracochlear implantation has the potential for playing a major role in the rehabilitation of certain deaf/blind individuals and that such patients may be amongst the most worthwhile to consider for cochlear implantation.

Case report

A 39-year-old blind man was admitted to our department because of profound deafness. He had suffered from bilateral otorrhoea since childhood following radical mastoidectomy. He lost hearing on the left side due to a labyrinthine fistula resulting from cholesteatoma. Hearing loss on the other side ensued as a result of an end-organ lesion as a consequence of labyrinthitis when he was 34 years old.

At the time of admission, pus and debris were found in both mastoid cavities despite antibiotic therapy and repeated aural toilet. The patient had skin lesions and a history of recurrent oral ulceration with respiratory and skin allergy. These abnormalities together with eye lesions resulting in blindness in his early childhood and the presence of the HLA-DR5 alloantigen, suggested the patient was suffering from Behçet's disease because of the presence of four of five diagnostic criteria of the disease according to suggestion of the International Study Group of Behçet's disease (1990). The laboratory findings showed normal blood glucose, electrolytes, haematology, and liver function tests. An audiogram demonstrated a profound hearing loss (Figure 1). No cochlear microphonic was observed, and there was no response to caloric stimulation with hot and cold air in either ear. The promontory test however, exhibited positive results ie. electrical excitability of the acoustic nerve was seen in both sides, subjectively confirmed by bilateral auditory sensation upon stimulation. Ultra high definition computed tomography (CT) scanning showed normal definition and a normal basal turn of the cochlea on both sides. Previously, he was rejected for cochlear implantation because of the otorrhoea possibly deriving from an incomplete removal of the secreting mucosa at previous mastoidectomy. In other respects he was considered a good candidate for a multichannel device.

Surgery

To overcome the problem of infected mastoid cavities he was submitted to two-stage surgery according to the description by Gray and Irving (1995). A post-auricular incision was made to approach the mastoid, the pinna was dissected and the external auditory canal was transected at the junction of bony and cartilaginous meatus. Fat was taken from the anterior abdomen. This was followed by a revision mastoidectomy. All residual middle-ear mucosa was removed together with the infected mastoid air cells. The mucosa of the Eustachian tube was also removed and its lateral wall was drilled away. The lumen was obstructed with muscle and bone paté. The bony cavity was then polished and a piece of silicone rubber sheeting was

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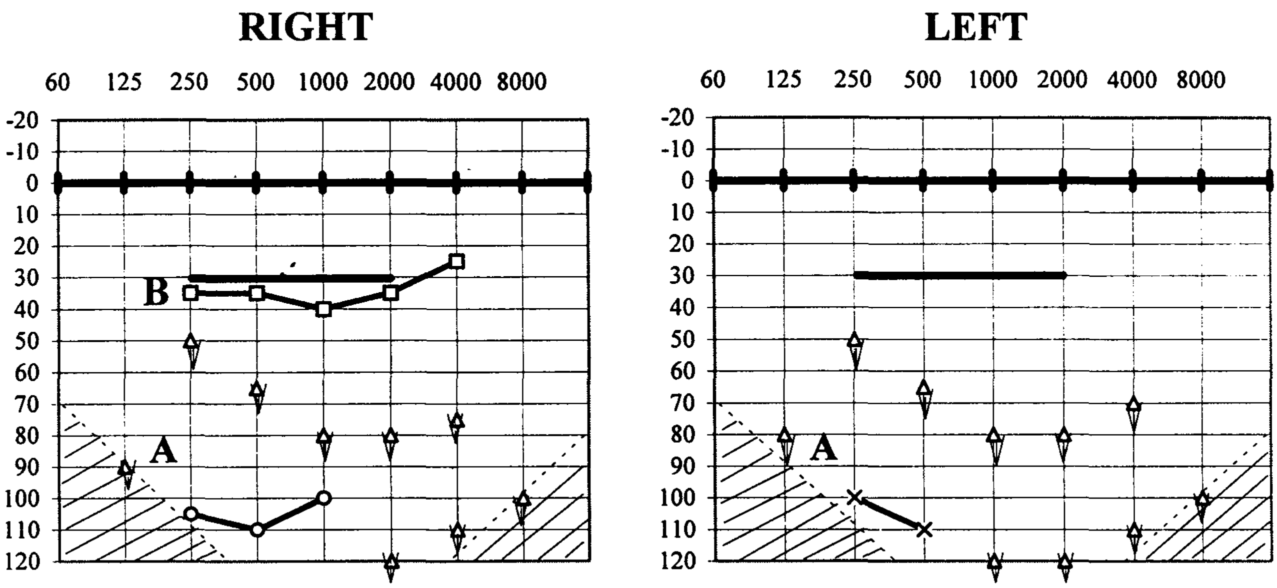

FIG. 1

Conventional audiology.
Curve A: Pre-operative unaided auditory thresholds. Curve B: Cochlear implant thresholds.

positioned over the promontory and round window. The cavity was obliterated with free abdominal fat. This was then followed by blind sac closure of the external auditory canal according to the description by Gray and Irving (1995).

Three months later, the ear was re-opened. The intended site of the implant was marked and a wide incision made. The site for the implant body was prepared. The fibrous fat was retracted forwards for access. The previously placed silicone disc was elevated and removed from over the round window and promontory. This was followed by cochleostomy and implantation of a Nucleus 22 ('Cochlear' GmbH Basel, Switzerland) multichannel device. All the 22 electrodes were positioned within the cochlea. The device was fixed near the round window. The wound was closed and the patient was discharged on the third post-operative day. One week after surgery, the correct intracochlear positions of the 22 electrodes were confirmed on a conventional transorbital radiograph. One month following implantation the device was activated by the audiologist.

Assessment of acoustic discrimination using the cochlear implant

We recorded cognitive responses as determined by the most commonly investigated components of cognitive event-related potentials such as the mismatch negativity (MMN), the N2b and the P300 to study the central auditory processes at the use of the implanted device. The study was performed using Dantec Concerto EEG-EP equipment (St Louis, MO) at an experimental setting of the so-called 'acoustic oddball paradigm'. The essence of this technique is that the patient is asked to calculate the number of target or deviant acoustic stimuli in a series of non-target or frequent stimuli. The acoustic oddball paradigm was used under passive conditions (the patient read a book for the blind over the investigation period) to verify the MNN and active conditions (the patient was asked to press a button after he had recognized the deviant stimuli) to confirm the other components (N2b and P300) (Regan, 1989; Oviatt and Kileny, 1991; Kraus *et al.*, 1992). The stimulation protocol included a 1000 Hz frequent and 2000 or 500 Hz deviant stimuli.

Results

Following switching on of the device, the patient acquired immediate open set speech discrimination ability and has continued to improve since then. As objectively indicated by results obtained from conventional audiology (Figure 1) and the auditory event-related potential study, the patient achieved an acoustic discrimination skill comparable to that seen in normally hearing subjects. Nevertheless, the implanted patient revealed a somewhat longer latency period (Figure 2a) with a smaller amplitude compared to that experienced by normally hearing individuals (Figure 2b).

Discussion

The results presented show that the deaf blind patient can benefit greatly from intracochlear implantation of a multichannel device. As indicated subjectively by immediate achievement of open set speech discrimination ability after switching on of the device (one month after surgery) and an excellent acoustic discrimination performance at the same time, the patient has regained his auditory communication skills with the outside world almost immediately following switch on at a baseline level.

The first deaf and blind adult patient to receive a Nucleus 22 multichannel intracochlear implant in Hungary underwent surgery in our department. He is the second deaf/blind Hungarian individual having an intracochlear implant and the first one with Nucleus 22 (the first deaf/blind Hungarian patient to receive an intracochlear implant is a child implanted with a Med-El device (Ribari *et al.*, 1997). This multihandicapped patient was of particular interest for several reasons. Primarily, because of his multi-sensory organ failure, secondly, since the profoundly deafened patient had been suffering from bilateral discharging ear for decades subsequent to radical mastoidectomy that took place in his childhood. Thus, the patient, otherwise suitable for cochlear implantation was rendered inappropriate for surgery because of the lack of a sterile, dry protected site for the electrodes. Thirdly, since he had a history of several gastrointestinal and respiratory disorders including gastric ulceration combined with gastrointestinal bleeding and respiratory allergy, we suspected a systemic disease.

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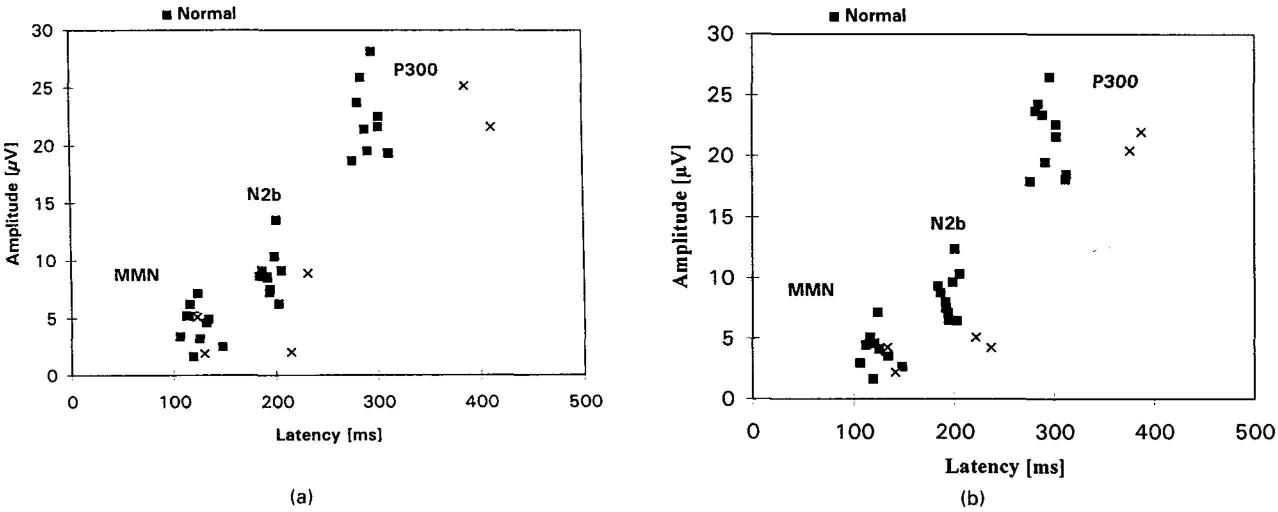

X values indicate individual data of two repetitive determinations with the implanted patient; the black squares denote two repetitive determinations with five normally hearing individuals. An amplitude latency relationship for mismatch negativity (MNN) (passive component and active components (N2b and P300).

a: 1000 Hz standard and 2000 Hz deviant stimuli; b: 1000 Hz standard and 500 Hz deviant stimuli.

FIG. 2

Auditory event-related potentials in a deaf/blind patient with a Nucleus 22 multichannel intraocochlear implant. A comparison with normally hearing subjects.

Cochlear implantation of deaf/blind individuals was previously thought risky bearing in mind the technique was thought to improve lipreading skills in normally sighted patients. This, technique at least in part, explains why only a relatively small number of deaf/blind have been implanted round the world. Futhermore, the risk of cochlear implantation is further amplified by the additional complication of chronic bilateral discharging ear excluding implantation without preceding pre-conditioning surgery. Indeed, our patient had also been refused intracochlear implantation because of the discharging ears at another implantation centre. We therefore decided to prepare the ear for cochlear implantation using the surgery proposed by Gray and Irving (1995). Subsequently, we found no difficulties in the implantation procedure.

It is difficult to state the reason why our patient showed such an excellent auditory performance immediately after device activation. According to the experiences of Ramsden *et al.* (1994) the deaf/blind have been amongst the very best performers. For an explanation, it is suggested that human individuals, similar to several animal species, who had been blind for years prior to the onset of deafness are better able to utilize auditory information than one might expect, because of adaptive plasticity of the cortex. The situation is different in patients who lose sight and hearing simultaneously or in those with preceding deafness. Whatever the precise explanation is, we think that deaf/blind patients with preceding blindness can derive greater benefit from multichannel intracochlear implantation than previously thought.

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