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Auditory Brainstem Implant in Postlingual Postmeningitic Patients

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Objectives/Hypothesis: The aim of this study was to evaluate outcomes of postlingual postmeningitic patients who received an auditory brainstem implant (ABI).

Study Design: Retrospective analysis was performed on postlingual postmeningitic patients with bilateral profound sensorineural hearing loss who underwent ABI between the years 2007 and 2014

Methods: All patients were postlingually deaf due to cochlear ossification as a consequence of bacterial meningitis. The patients received a MED-EL or Neurelec ABI. All patients were operated on at different hospitals by the same primary surgeon. The patients were tested using Ling 5 sound detection, sound field implant thresholds between 250 Hz and 6 kHz, and 6 to 12 choice closed-set word and sentence tests.

Results: Nine patients with postmeningitic cochlear ossification received an ABI. Five of nine ABI users (55.5%) wear their audio processors (AP) most of the time. Four (44.5%) with no perceivable benefit have become nonusers. Three of the five consistent ABI users reported good benefit. The other two ABI users who do wear their APs do not respond to sound in daily living but reported benefits such as "feeling sound" in a good way.

Conclusions: In this study, five of nine patients (55.5%) with bilateral ossified cochlea had some degree of benefit from their ABI. An ABI may be useful in hearing restoration in postlingual patients with bilateral ossified cochlea due to meningitis. However, poor results may be related to side effects, which may necessitate deactivation of electrodes, long duration of auditory deprivation, or impairments in the auditory neural structures as a result of meningitis.

Key Words: Auditory brainstem implant, meningitis, cochlear ossification.

Level of Evidence: 4.

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INTRODUCTION

Bacterial meningitis is one of the main reasons of cochlear ossification that result in profound hearing loss. In some patients, cochlear ossification does not involve the entire cochlea, and cochlear implantation may provide adequate auditory rehabilitation in such cases.[1] However, the reported results of cochlear implantation are not satisfactory in severe cochlear ossification, although many techniques have been proposed to deal with this problem.[2,3] The poor results of cochlear implants in patients with severe cochlear ossification have been attributed to peripheral nerve degeneration,[4] unstable insertion of electrodes, insufficient contact of electrodes to the spiral ganglion, and decreased number of viable auditory neurons.[5–9] Accordingly, it is difficult and sometimes impossible to perform cochlear implantation in patients with severe cochlear ossification. An auditory brainstem implant (ABI) can provide auditory rehabilitation in situations where cochlear implantation is not possible.[7]

The aim of this study was to evaluate outcomes of postlingual postmeningitic patients who received an ABI.

MATERIALS AND METHODS

This retrospective study included patients with bilateral profound sensorineural hearing loss who underwent auditory brainstem implantation between the years 2007 and 2014. All patients were postlingually deaf due to cochlear ossification as a consequence of bacterial meningitis. All patients were operated on at different hospitals by the same primary surgeon.

The ABI surgeries were performed via a retrosigmoid approach. During the surgery a 3 × 3-cm craniotomy was made in the retrosigmoid area. The cerebellum was retracted after releasing the cerebrospinal fluid in the cisterna magna. The arachnoid and its adhesions were dissected. The foramen Luschka was identified between the root of the ninth cranial nerve and choroid plexus after dura incision and cerebellum retraction. The ABI electrode was placed on the cochlear nucleus in the foramen Luschka.

All MED-EL users were fitted and followed up by the same audiologist. The Neurelec user was fitted and followed up by a different audiologist. Initial switch-on was performed with the ABI user awake but under cardiac monitoring in the operating room. At initial stimulation, after establishing that the charge level used to measure impedance field telemetry (IFT) was not causing the ABI user any discomfort, telemetry

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TABLE I.
Characteristics of Patients.

Patients	Age (Years)	CI Trial	Length of Hearing Deprivation (Years)	Type of Implant and Processor	Length of ABI Use (Months)
1	46	Yes	10	Concerto Opus 2	31
2	40	No	19	Pulsar Tempo+	40
3	23	Yes	10	Concerto Opus 2	33
4	28	Yes	13	Concerto Opus 2	29
5	30	No	8	Pulsar Tempo+	93
6	46	No	29	Concerto Opus 2	29
7	47	Yes	1.5	Concerto Opus 2	69
8	33	Yes	1.5	Concerto Opus 2	10
9	17	Yes	10	Neurelec Digisonic SP	6

CI = cochlear implant, ABI = auditory brainstem implant.

measures were made and evaluated. At switch-on and follow-up fits, threshold (THR) levels were measured on all active electrodes. If users had hearing as opposed to a side effect, such as leg tingling or constriction in the throat, then stimulation was slowly increased above the THR level until the user reported the sound to be loud or until they reported that a side effect had started and was causing them discomfort. Any electrodes resulting in some hearing with some dynamic range (charge difference between THR and maximum comfort level [MCL]) and no side effect were left activated. Electrodes where stimulation did not result in hearing but only side effects were deactivated. Once a program was configured and switched on (live mode), MCLs were globally increased until the user reported that sound was too loud or a side effect had started. Each ABI user's latest audio processor (AP) program was analyzed, with the status of electrodes as defined by IFT; mean charge levels, number of active electrodes, and most prevalent side effects were recorded.

Latest reports obtained from each ABI user were analyzed. AP wearing habits and the ABI users' perceived benefit from their ABIs were recorded. Performance of ABI users with their latest AP program was checked. ABI users with no hearing could not be tested. Tests included: Ling 5 sound (/a/, /ee/, /u/, /sh/, and /s/) detection, sounds were presented with "live" voice at quiet conversational level, sound field implant THRs between 250 Hz and 6KHz (conducted in a sound-proofed cabin), and six to 12 choice closed-set word and sentence tests. The closed-set word tests used were the mono, trochee, polysyllabic test and the bisyllabic word test from the MED-EL EARS test battery adapted into the Turkish language. Sentences used for closed-set testing were chosen together with the ABI user, written down, and practiced with auditory and visual cues, before sentence recognition was tested using live voice and auditory cues only.

RESULTS

Nine patients with postmeningitic cochlear ossification received an ABI. The mean age at implantation of ABI users was 34.4 years, ranging from 17 to 47 years. There were seven (77.7%) male and two (22.3%) female patients. All ABI users had bilateral ossified cochleae as a sequelae of bacterial meningitis. In six patients a cochlear implantation was attempted but no implantation was made because of total ossification of the cochlea. In the remaining patients no prior cochlear implantation was attempted because a complete ossification of the cochlea was confirmed by computed tomography and magnetic resonance imaging. The mean duration of sound deprivation was 11.3 years, ranging from 18 months to 29 years.

Eight patients received a MED-EL and one patient received a Neurelec ABI. Seven patients (77.7%) were implanted on the right side and two patients (22.3%) were implanted on the left side. The mean duration of ABI use for eight users was 30.8 months (range, 6–69 months). One ABI user has been using his ABI for nearly eight years. Of 8 MED-EL ABI users, six had a CONCERTO ABI and an OPUS 2 AP and two had a PULSAR ABI and used a TEMPO+ AP. The Neurelec user had a Digisonic SP ABI. Characteristics of patients are summarized in Table I.

Latest IFT measures for MED-EL ABI users were normal with impedances ranging from 2.00 to 9.56 KΩ. No electrodes had high impedances or short circuits. All ground path impedance values were normal, ranging from 0.37 to 1.22 KΩ. The mean MCL charge level for all eight MED-EL ABI users was 147 qu but for the three better performers was 78 qu. The average number of active electrodes was 4.25 for all eight MED-EL ABI users, but 6.3 for the three better performers. Most frequently activated electrodes were electrode (E) 5 and E7, followed by E4 and E10, the least frequently activated electrodes were E6 and E8. The most commonly seen side effects were constriction to throat and leg tingling.

Five of nine ABI users (55.5%) wear their APs most of the time. Three of the five consistent ABI users reported good benefit, and this is reflected in their performance. The other two ABI users who did wear their APs did not respond to sound in daily living but reported benefits such as "feeling sound" in a good way and relief from headache. Three ABI users who clearly benefit from their ABIs had sound field THRs varying from 25 to 50 dB HL. Sound field THRs were also measurable for patient 6 who had poorer high-frequency responses. Table II shows ABI sound field thresholds from 250 Hz to 6 kHz for patients 3 to 6. These users had access to quiet conversational speech. Four (44.5%) patients have become nonusers. Three of them had no perceivable benefit. The patient who received Neurelec ABI had limited benefit. He could detect two out of six Ling sounds. However, he was uncooperative during speech perception testing. Unfortunately, he refused to wear the external unit and has become a nonuser.

TABLE II.
Auditory Brainstem Implant Sound Field Thresholds From 250 Hz-6 kHz for Patients 3 to 6.

	250 Hz	500 Hz	1 kHz	2 kHz	4 kHz	6 kHz
Patient 3	45	45	45	35	50	40
Patient 4	30	30	45	30	30	25
Patient 5	30	30	35	35	45	35
Patient 6	40	25	40	55	75	NR

NR = no response.

TABLE III.
Postoperative Results of Auditory Brainstem Implant.

Patients	Telemetry and GPI (KΩ)	Mean Charge (qu)	Active Electrodes	Benefit	Ling Sound Detection	Sound Field Implant Threshold (dB HL) 0.25–6 kHz	Closed-Set Word Score (%)	Closed-Set Sentence Score (%)	Wearing Habit
1	All OK 2.00–7.23, GPI 0.37	162	5, 7, 10	No	—	—	—	—	Become nonuser
2	All OK 3.16–6.47, GPI 0.99	237	9, 11, 12	No	—	—	—	—	Become nonuser
3	All OK 3.55–7.30, GPI 0.55	95	10, 9, 7, 5, 4, 3, 2, 1	Significant benefit	5 of 5	45, 45, 45, 35, 50, 45	MTP 92% BIS 67%	90%	Always wears
4	All OK 4.79–9.56, GPI 0.45	56	10, 7, 2, 5, 4	Significant benefit	5 of 5	30, 30, 45, 30, 30, 25	BIS 71%	80%	Always wears
5	All OK 2.71–7.74, GPI 1.22	84	4, 5, 7, 9, 10, 12	Significant benefit	5 of 5	30, 30, 35, 35, 45, 35	MTP 50%	70%	Always wears
6	All OK 3.18–7.80, GPI 0.64	237	2, 4, 6, 1, 3	Limited benefit	4 of 5	40, 25, 40, 55, 75, NR	MTP 100%	4 choice 100%	Always wears
7	All OK 5.00–7.50, GPI 0.53	69	8, 11	No	—	—	—	—	Become nonuser
8	All OK 0.62–3.60, GPI 0.62	237	5, 7	Limited benefit	—	—	—	—	Always wears
9	All OK	230	5, 15	Limited benefit	2 of 6	—	—	—	Become nonuser*

*Refused to wear an external unit although he had some benefit.
BIS = bisyllabic; GPI = ground path impedance; MTP = mono, trochee, polysyllabic; NR = no response.

The three higher performers could detect five out of five Ling sounds spoken softly; patient 6 could detect four out of five Ling sounds (not /s/) in the test situation. Patient 6, however, was not able to spontaneously respond to sound in daily living. The better performers, patients 3, 4, and 5, scored between 50% to 67% on 12 choice closed-set word recognition tests and 70% to 90% on 10 choice closed-set sentence recognition tests. Patient 6 could score above chance level on six choice closed-set word recognition and four choice closed-set sentence recognition tests. None of the patients were able to do open-set testing. Table III summarizes the results and parameters of ABI.

DISCUSSION

In severe cochlear ossification, cochlear implantation may be very difficult or impossible.[10] Several techniques, including scala vestibuli insertion,[11] multiarray electrodes,[12,13] insertion through middle cranial fossa,[14] and basal turn drill-out,[3] have been proposed to overcome cochlear ossification. However, these techniques usually yield suboptimal or no results.

The outcomes of cochlear implantation in postmeningitic deafness are variable. Degree of ossification is the most important factor affecting auditory performance after cochlear implantation. Rauch et al.[3] reported poor auditory results in four patients who underwent radical drill-out for severe labyrinthitis ossificans. Only 17% of cases with complete ossification had open-set sentence recognition compared with 83% of cases with nonossified cochleae and 38% of cases with partial ossification. Steenerson and Gary[15] reported results of cochlear implantation for postmeningitic deafness. They compared children who required drill-out with those who did not require drill-out. Test performance for speech understanding was highest in the nonossified group and lowest in the ossified group with complete drill-out. However, children with extensive ossification requiring complete drill-out had some benefit from cochlear implantation. El-Kashlan et al.[16] evaluated the effects of degree of ossification on auditory performance in prelingually deaf children who underwent cochlear implantation, and found that children with cochlear ossification had a lower speech perception category than those with nonossified cochleae. The authors also reported that four of the children with cochlear ossification demonstrated open-set speech recognition with long-term implant use. Nichani et al.[1] compared auditory performance outcomes in ossified and nonossified cochleae among children who underwent cochlear implantation after bacterial meningitis, and found that children with postmeningitic deafness benefit significantly from cochlear implantation. Rotteveel et al.[17] evaluated the long-term outcome of children with postmeningitic deafness and compared speech perception performance of partial-insertion and full-insertion cases. They found that speech perception in the partial-insertion children was poorer than that in the control groups. However, four of seven children with partial insertion acquired open-set word recognition. The authors concluded that the patients with partial insertion of the electrode array might have some benefit from a cochlear implant. Cohen and Waltzman[18] reported poor speech perception outcomes in most of their partial insertion cases. De Barros et al.[19] reported that four out of five patients who underwent cochlear implantation for postmeningitic deafness had good results.

Satisfactory results were reported with ABI in patients with severe cochlear ossification. Grayeli et al.[5] first reported a case of postmeningitic deafness with totally ossified cochlea. Twelve electrodes were activated. Fifty percent disyllabic word scores and 60% sentence scores were achieved. These scores reached 80% and 93%, respectively, with lip reading. In a later study, Grayeli et al.[20] reported three cases of postmeningitic profound hearing loss with complete cochlear ossification. All patients had significant speech discrimination in sound-only mode and enhanced lip reading performance with an ABI. Sanna et al.[2] performed ABI on a 12-year-old female child with postmeningitic deafness and bilaterally ossified cochleae. The authors reported that the patient can freely use the telephone after 8 months of implantation. Colletti et al.[7] performed ABI on three adults after unsuccessful cochlear implantation, and the patients had discrimination of two- or three-syllable words with scores from 85% to 100%. Choi et al.[6] reported three patients with ossified cochlea who underwent ABI. All patients reported auditory sensations when the ABI was activated. One patient achieved open-set speech recognition and had no difficulty with communication. A second patient had improved closed-set speech discrimination with visual cues but only limited open-set recognition scores. A third patient was not able to use the device due to nonauditory stimulation.

In this cohort, five of nine ABI users had some degree of benefit from their ABI. Although most of the ABI users could achieve satisfactory closed-set recognition, none of the patients were able to achieve open-set speech discrimination. Three users received significant benefit allowing them to converse quite freely with conversational partners as long as they could see the speakers face. Their ABIs also provided them with enough access to sound to enable them to understand some words and sentences through audition only, when they are in quiet surroundings and are cued into the "topic."

In the current study, even though all nine ABIs are fully functioning, the benefit users receive from them is limited. One reason for this limited benefit may be the long mean length of sound deprivation users experienced prior to implantation. A more likely reason, however for limited benefit, is the high prevalence of side effects. Side effects prevent sufficient charge being delivered to bring about hearing. Two users, patients 3 and 8, in this cohort had such severe side effects that it was not possible for them to use their ABI. These side effects and poor performance may be related to adhesions and scarring in the neural tissues as well as auditory pathways that are possible after bacterial meningitis. Similar adhesions and scarring also apply to the arachnoid membrane, which usually has a ground glass appearance and needs meticulous dissection during surgery of postmeningitic patients. The better performers in this cohort required smaller charge levels to hear and had more active electrodes. Smaller charge requirements and more active electrodes are indicators of better performance.

CONCLUSION

Five of nine patients (55.5%) with bilateral ossified cochlea had benefit to some degree from their ABI. Three users received significant benefit allowing them to converse quite freely with conversational partners as long as they could see the speakers face. ABI may be useful in hearing restoration in postlingual postmeningitic patients with bilateral ossified cochlea. However, poor results may be related to side effects, which may necessitate deactivation of electrodes, long duration of auditory deprivation, or impairments in the auditory neural structures as a result of meningitis.

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