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Cochlear Implantation in a Patient with Mitochondrial Encephalopathy, Lactic Acidosis and Stroke-like Episodes Syndrome

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Yasumura S, Aso S, Fujisaka M, Watanabe Y. Cochlear implantation in a patient with mitochondrial encephalopathy, lactic acidosis and stroke-like episodes syndrome. *Acta Otolaryngol* 2003; 123: 55–58.

Objective—The aim of this study was to investigate the usefulness of a Nucleus® CI24M cochlear implant in a patient with mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS) syndrome.

Material and methods—A 29-year-old woman with MELAS syndrome presented with sensorineural hearing loss and diabetes mellitus and underwent multichannel cochlear implantation. The follow-up period was 10 months. The case history, assessment of mitochondrial disease, indications for the cochlear implant and the benefits of cochlear implantation were evaluated.

Results—Nine months after the surgery the patient could use the telephone and was quite satisfied with the improvement in communication due to the cochlear implant.

Conclusion—Cochlear implantation can be recommended for patients with MELAS syndrome if they have residual retrocochlear function. Single photon emission computerized tomography was found to be very useful for evaluating retrocochlear function. *Key words:* central nervous system, retrocochlear function, single photon emission computerized tomography.

INTRODUCTION

Mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS) syndrome is one of the mitochondrial encephalomyopathies. As the name suggests, the main features are mitochondrial myelopathy, encephalopathy, lactic acidosis and stroke-like episodes. Additional symptoms include fatigue, muscle weakness, sensorineural hearing loss (SNHL), cardiomyopathy, diabetes mellitus and mental disorders. The most common physical characteristic of patients with MELAS syndrome is low stature. Hearing loss is observed in a high proportion of patients who develop clinical features of MELAS (1). The lesion responsible for hearing loss can be associated with both cochlear and retrocochlear involvement. Recently, cochlear implantation has been widely used to help patients with bilateral profound or total deafness resulting from a variety of congenital and acquired causes. However, it remains controversial as to whether patients who become deaf as a result of encephalopathy including MELAS syndrome should undergo cochlear implantation. In this study, we report a case of MELAS syndrome in which a cochlear implant improved the sound awareness, speech recognition and communication ability of the patient. Brain single photon emission computerized tomography (SPECT) was useful with regard to deciding the indication. Cochlear implantation can improve a patient's quality of life, even when hearing loss has progressed and is combined with other disorders.

CASE REPORT

The subject was a 29-year-old woman diagnosed as having SNHL at the age of 13 years. She had normal prenatal and perinatal histories. Her family history was also normal. Diabetes mellitus was noted at the age of 15 years and insulin therapy was initiated at 19 years. At the age of 22 years, her hearing acuity was noted to be severely impaired: 90 dB in the right ear and 110 dB in the left. She noted that her hearing aid was no longer of benefit. One year later, she reported dizziness and weakness in her right arm and foot. Serum biochemical studies showed a high level of lactic acidosis, which increased after exercise. Assessment for mitochondrial disease detected the mitochondrial DNA (mtDNA) 3243 mutation and treatment with coenzyme Q was started. Lactic acidosis improved, but hearing loss and tinnitus did not change after the treatment. At the age of 24 years, she experienced double vision, visual field defects, dizziness, nausea, consciousness disorder, headaches and a deterioration in hearing loss. The visual symptoms and consciousness disorder resolved spontaneously, but the other symptoms persisted. The mtDNA 3243 mutation and symptoms confirmed a clinical diagnosis of MELAS syndrome. Five years later, her bilateral hearing had gradually worsened and she had begun to communicate using lip reading and writing. Her speech production was well preserved.

At the age of 29 years, she was admitted to our hospital for cochlear implantation. Preoperative pure-

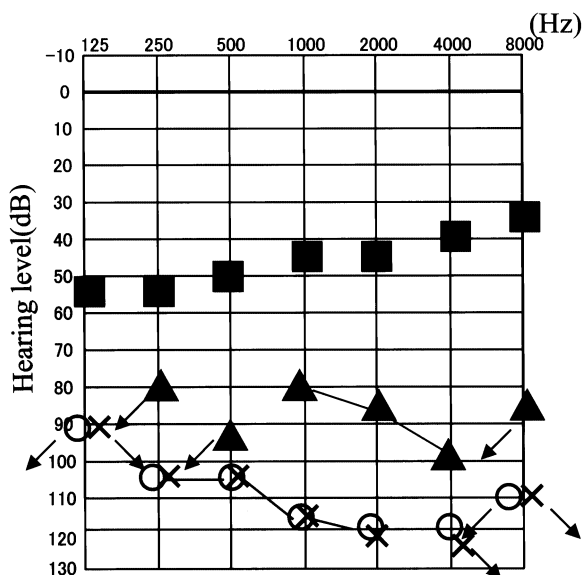


Fig. 1. Audiograms before and after cochlear implantation: (○) right ear hearing level; (×) left ear hearing level; (▲) with hearing aid (sound-field threshold); (■) with cochlear implant (sound-field threshold).

tone audiometry revealed bilateral profound hearing loss with an average of 114 dB in both ears (Fig. 1). The hearing aid was no longer useful. Speech recognition scores were measured using a Japanese word list. Her sentence recognition scores were 0% and 4% in open- and closed-set listening, respectively (Fig. 2). Promontory electrocochleography and brainstem auditory-evoked potentials (BAEPs) showed no re-

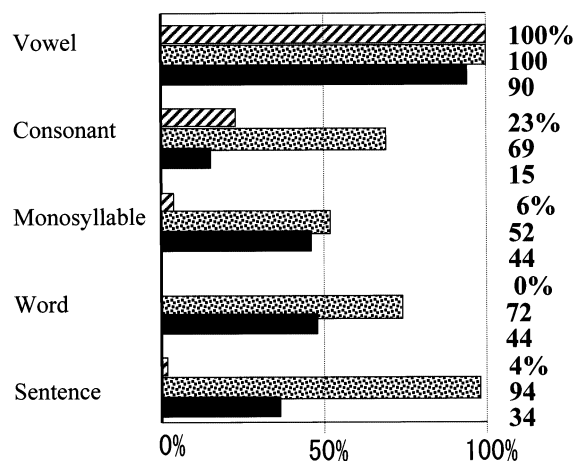


Fig. 2. Pre- and postoperative speech recognition scores observed using a Japanese word list. Vowels and consonants were tested using a live voice and other sounds were tested using a videotape that is widely used in cochlear implantation centers in Japan. Note that preoperative open-set scores were 0% for all the recognition tasks (data not shown). *Hatched bars*: closed-set listening before surgery; *stippled bars*: closed-set listening after surgery; *filled bars*: open-set listening after surgery.

Table I. Results of the promontory stimulation test

	100 Hz bursts (μA)	200 Hz bursts (μA)
Right ear		
Threshold	10.3	14.1
Maximum acceptable loudness	19.3	24.5
Dynamic range	9.0	10.4
Left ear		
Threshold	10.2	15.8
Maximum acceptable loudness	18.4	34.4
Dynamic range	8.2	18.6

Average stimulation models from two trials are shown. The patient could discriminate the frequency difference between 100 and 200 Hz.

sponse in either ear. In a promontory stimulation test, the patient responded to electrical stimulation in both ears (Table I). She could discriminate the frequency differences. Gap detection tests and temporal difference limen tests could not be performed because the patient felt dizzy and had a headache during the tests. Both tympanic membranes were normal. CT and MRI scans showed no abnormality in either inner ear (Fig. 3). MRI demonstrated cerebral atrophy. Although the brain showed a low uptake of ^{99m}Tc in the occipital area, perfusion in the left temporal area was not significantly decreased (Fig. 4).

On 6 March, 2000 she underwent cochlear implantation surgery using a 24-channel device (Nucleus® CI24M) in the right ear. Twenty-five electrode rings were successfully inserted and all 22 channels and 2 extracochlear electrodes were found to be usable at the initial “switch on” of the cochlear implant. Three months later, speech recognition scores had improved as follows: word recognition score, 0% with the hear-

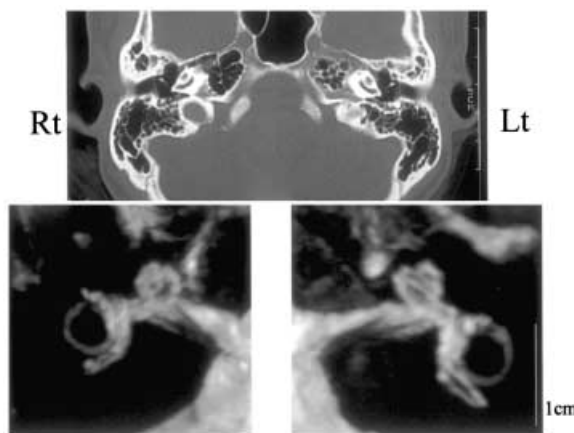


Fig. 3. CT and MRI scans of both inner ears. The three-dimensional Fourier transform contractive interference in steady state technique was adopted for the MRI scan.

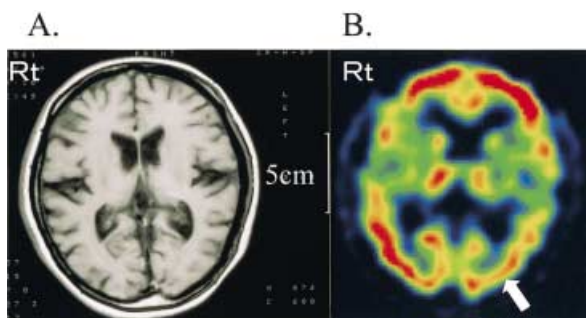


Fig. 4. (A) Brain MRI scan (T1 image) demonstrating cerebral atrophy. (B) Brain SPECT scan (axial image) showing slightly low uptake (arrow) of ^{99m}Tc in the occipital area. Perfusion in the left temporal area was not significantly decreased.

ing aid to 72% with the cochlear implant and sentence recognition score, 4% to 94% in closed-set listening; word recognition score, 0% to 44% and sentence recognition score, 0% to 34% in open-set listening (Fig. 2). Postoperative neural response telemetry (NRT), implant-evoked BAEP and middle latency response (MLR) showed good responses, as shown in Fig. 5. Nine months after the surgery, the patient could use the telephone and was quite satisfied with the improvement in communication due to the cochlear implant.

DISCUSSION

Mitochondrial encephalomyopathy is the disorder associated with abnormal mitochondrial function. The disease is maternally inherited and is often observed among family members. Based on its clinical features, it is classified into 3 types: (i) chronic external ophthalmoplegia; (ii) MELAS syndrome; and (iii) myoclonic epilepsy associated with ragged red fibers. Many cases of mitochondrial encephalomyopathy are associated with mtDNA mutations and mtDNA 3243 and mtDNA 3271 mutations have been detected in

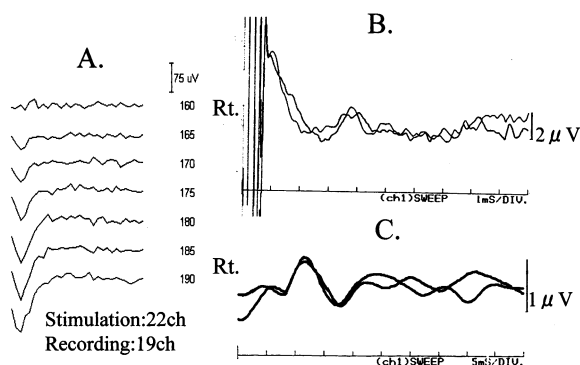


Fig. 5. (A) Postoperative NRT, (B) implant-evoked BAEP and (C) MLR results.

80% and 10% of patients with MELAS syndrome, respectively (2). Our patient had an mtDNA 3243 mutation but no mutation of mtDNA was observed and MELAS syndrome did not occur in her mother.

It is known that SNHL is associated with MELAS syndrome. It was previously reported that SNHL is observed in 27–47.6% of patients with MELAS syndrome (1, 3). Hearing loss occurs between the ages of 14 and 50 years. Most patients have bilateral progressive SNHL. The shape of the audiogram is generally sloping at the beginning, and gradually becomes flat (4). Clinically, a decrease in speech acuity and auditory agnosia are occasionally observed in the advanced stage. Speech discrimination scores are generally poor and do not parallel the degree of hearing loss. The lesion responsible for hearing loss can be associated with both cochlear and retrocochlear involvement. (4). Preoperative auditory assessment, promontory stimulation test, electrically-evoked BAEPs and MLR should be considered as useful tools in order to select patients with MELAS syndrome for cochlear implantation. As the promontory stimulation test was positive, a severe central nervous disorder was not observed and a brain SPECT scan showed temporal uptake in our patient, she was considered suitable for cochlear implantation. Positron emission tomography and SPECT were also useful for preoperative patient selection. SPECT was considered to be very useful as it could be performed more easily and less expensively than PET.

The histopathological features of the inner ear in patients with MELAS syndrome have not yet been described. Lindsay and Hinojosa (5) reported the histopathological features of a patient with Kearns–Sayre syndrome to be as follows: advanced cochleolaccular degeneration, including the collapse of Reissner's membrane; degeneration of the organ of Corti, tectorial membrane and stria vascularis; a considerable reduction in the number of spiral ganglion cells; and the degeneration of nerve fibers within the osseous spiral lamina. Pathological changes in the central nervous system (CNS) of patients with MELAS syndrome have been reported to be as follows: diffuse atrophy of the cerebral and cerebellar cortices; diffuse gliosis of the cerebral and cerebellar white matter; and cactus formation of Purkinje cells (6). These findings suggest that both cochlear and retrocochlear pathological changes may be caused by mitochondrial disorders. Therefore, lesions originating in the inner ears may reach the retrocochlear and auditory cortices as the disease progresses in patients with MELAS syndrome.

Our patient obtained good implant-evoked responses and excellent hearing performance following cochlear implantation. However, the indication of cochlear implantation for MELAS syndrome is still

controversial because it is a progressive disease and patients will lose hearing acuity in the future. The progression of MELAS syndrome is relatively slow and the long-term prognosis is reasonable. Rosenthal et al. (7) and Sue et al. (8) reported successful outcomes in MELAS syndrome patients with significant CNS degeneration who underwent cochlear implantation. We found that cochlear implantation in a patient with multisystem degenerative disease greatly contributed to the patient's quality of life and made it possible for her to communicate with family members and caregivers better than previously.

CONCLUSION

We have described successful cochlear implantation in a MELAS syndrome patient with CNS lesions. The results indicated that cochlear implantation can be recommended for patients with MELAS syndrome if they have residual retrocochlear function. SPECT was considered to be useful for preoperative patient selection.

REFERENCES

1. Pavlakis SG, Phillips PC, Dimauro S, et al. Mitochondrial myopathy, encephalopathy, lactic acidosis, and strokelike episodes; a distinctive clinical syndrome. *Ann Neurol* 1984; 16: 481–8.
2. Goto Y, Horai S, Matsuoka T, et al. Mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episode (MELAS): a correlative study of the clinical features and mitochondrial DNA mutation. *Neurology* 1992; 42: 545–50.
3. Elverland HH, Torbergesen T. Audiologic findings in a family with mitochondrial disorder. *Am J Otol* 1991; 12: 459–65.
4. Tamagawa Y, Kitamura K, Hagiwara H, et al. Audiologic findings in patients with a point mutation at nucleotide 3243 of mitochondrial DNA. *Ann Otol Rhinol Laryngol* 1997; 106: 338–42.
5. Lindsay JR, Hinojosa R. Histopathologic features of the inner ear associated with Kearn–Sayre syndrome. *Arch Otolaryngol* 1976; 102: 747–52.
6. Tanahashi C, Nakayama A, Yoshida M, Ito M, Mori N, Hashizume Y. MELAS with the mitochondrial DNA 3243 point mutation: a neuropathological study. *Acta Neuropathol (Berl)* 2000; 99: 31–8.
7. Rosenthal EL, Kileny PR, Boerst A, Telian A. Successful cochlear implantation in a patient with MELAS syndrome. *Am J Otol* 1999; 20: 187–91.
8. Sue CM, Lipsett LJ, Crimmins DS, et al. Cochlear origin of hearing loss in MELAS syndrome. *Ann Neurol* 1998; 43: 350–9.

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