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Cochlear Implantation via a Transmeatal Approach in an Adolescent with Type II Mucopolysaccharidosis (Hunter Syndrome): A Case Report

Yun-Hoon Choung[1]*, Hantai Kim[2], Jun Young An[2], Oak-Sung Choo[2] and Jeong Hun Jang[3]

[1]*Department of Otolaryngology, Ajou University School of Medicine, Korea*

[2]*Department of Medical Sciences, Ajou University Graduate School of Medicine, Korea*

[3]*BK21 Plus Research Center for Biomedical Sciences, Ajou University Graduate School of Medicine, Republic of Korea*

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***Corresponding author:** Yun-Hoon Choung, Department of Otolaryngology, Ajou University School of Medicine, Republic of Korea, Korea

Abstract

Type II mucopolysaccharidosis (MPS II), or so-called Hunter syndrome, is a rare X-linked lysosomal storage disorder caused by an enzyme deficiency of iduronate-2-sulfatase, presenting otorhinolaryngological manifestations including sensorineural hearing loss (SNHL). Previously, the median survival age of MPS patients was approximately 13.4 years. However, after the introduction of enzyme replacement therapy and other multidisciplinary care modalities, the life expectancy has risen. Thus, surgeons will encounter more, relatively older, MPS patients with progressive SNHL. Here, we report the rare case of an adolescent with MPS II who underwent cochlear implantation (CI) for the treatment of severe SNHL. We report a case of a 13-year-old adolescent with MPS II. Due to unpredicted mastoid emissary veins and overgrowth of vessels around the temporal bone, CI was performed via transmeatal approach, not by conventional transmastoid method, to prevent post-operative bleeding. Of the 20 electrodes, 18 were inserted through a round window, and the inserted electrodes were protected with cartilage without external canal obliteration. There was no substantial bleeding or other intra-operative complications. The patient was discharged without any problems, two days after the operation. Average of post-operative hearing thresholds was 35 dB and none of surgical complications have been reported. CI is admitted as a considerable option for MPS II with severe SNHL. However, in contrast to the CI at the early age in the former studies, adolescent MPS II may present unpredicted anatomical abnormalities, such as the vessel overgrowth, which would restrict successful performance of the operation. Therefore, a surgeon should deliberately evaluate the anatomical abnormalities before a surgery.

Keywords: Cochlear Implantation; Mucopolysaccharidosis II; Hunter Syndrome; Sensorineural Hearing Loss

Abbreviations: MPS: Mucopolysaccharidosis; GAGs: Glycosaminoglycans; MPS II: Type II Mucopolysaccharidosis; SNHL: Sensorineural Hearing Loss; ERT: Enzyme Replacement Therapy; CI: Cochlear Implantation; MRI: Magnetic Resonance Imaging; CT: Computed Tomography; EAC: External Auditory Canal

Introduction

Mucopolysaccharidosis (MPS) is a rare inherited disorder characterized by the degradation of glycosaminoglycans (GAGs) [1]. Excessive accumulation of GAGs in tissues alters cellular functioning with various clinical consequences [1,2]. Type II MPS (MPS II), or Hunter syndrome, is a rare X-linked lysosomal storage disorder caused by a deficiency in iduronate-2-sulfatase, presenting with a coarse facies, hepatomegaly, airway and pulmonary dysfunction, skeletal deformities, neurological impairment, heart problems, adenotonsillar hypertrophy, and otitis media [3]. And, sensorineural hearing loss (SNHL) is represented in 7% of patients [4]. Enzyme replacement therapy (ERT) is the usual treatment. Recombinant human iduronate sulfatase, termed idursulfase, is administered weekly [5]. However, supportive or symptomatic management is also important. Particularly, when SNHL is present, otologists can improve the quality of life for patients and their families. Currently, cochlear implantation (CI) is an established method for the treatment of severe SNHL. Saeed et al. found and reported that CI was a useful therapeutic option for MPS II with SNHL. In the report, a male patient underwent CI aged 4 years. No potential anatomical obstacles for CI had not been identified in CT scanning [6]. Previously, the median survival age of MPS patients was approximately 13.4 years [7]. However, after the introduction of ERT and other multidisciplinary care modalities,

the life expectancy has risen. Thus, surgeons will encounter more, relatively older, MPS patients with progressive SNHL. Here, we report the rare case of an adolescent with MPS II who underwent CI for the treatment of severe SNHL.

Case Report

A 13-year-old Korean male adolescent with MPS II was referred because of progressive bilateral hearing impairment. He exhibited valvular cardiopathy, a bilateral equinocavus deformity, kyphosis, a developmental language delay, and progressive bilateral hearing impairment. He had been taking idursulfase since 2009. He had used bilateral hearing aids since the age of 10 years. The hearing thresholds on pure-tone audiometry were 75 dB in both ears (Figure 1), and the speech discrimination scores were 44% at 90 dB in the right ear and 40% at 90 dB in the left ear. This score had improved by up to 56% at 65 dB in both ears with the use of bilateral hearing aids. Brain stem audiometry revealed hearing thresholds of 70 dB in the right ear and 75 dB in the left ear. A vestibular function evaluation revealed no abnormality. The inner ear structures were relatively normal on cochlear magnetic resonance imaging (MRI) and high-resolution temporal bone computed tomography (CT). Both mastoid cavities were underdeveloped and exhibited poor pneumatization. Suspicious emissary veins were evident in the bilateral portion of the mastoid (Figures 2A-2C). Both cochlear nerves were apparent on oblique, sagittal cochlear MRI.

The first operation used a conventional transmastoid approach in the left ear. However, both the mastoid emissary veins and overgrowth of vessels around the temporal bone limited surgical progress. Eventually, the operation was terminated by ligating the vessels and applying bone wax (Figures 3A-3D). We considered several issues when preparing for the second operation. First, we evaluated the arterial blood vessels via temporal bone CT with angiographic three-dimensional reconstruction; this revealed vessel overgrowth. Fortunately, the overgrowth did not compromise the desired surgical approach. Second, we considered the implantation site. As venous vessel development in the right ear was unusual, we thought that CI in the left ear should be performed using a retroauricular transmeatal approach to avoid emissary veins. And, we also thought that CI device with screw type was the best option because it was not necessary to drill out for seating an internal part. Third, we contemplated the length of the electrode array. For example, a 60–70-mm array was too short; we could not deliver the electrodes from the receiver-stimulator via a transmeatal approach. Also, emissary veins were present where the receiver-stimulator would usually be placed. Thus, we chose to place an array that was 80–90 mm in length of Neuro Zti Implant (Oticon Medical Systems, Vallauris, France).

CI was performed in the left ear 4 months later. The vessel ligation performed during the first surgery eliminated substantial bleeding. After dissection of the skin of the external auditory canal (EAC), the posterior EAC wall was drilled to form a groove for the electrode array. A pseudo membrane around the round window was identified and removed, and electrodes inserted through the round window. The electrode insertion site was covered with cartilage. Of the 20 electrodes, 18 were inserted and responded to intraoperative neural response telemetry. The electrodes were fixed with Greenplast and surgical bone wax. We encountered no intraoperative or postoperative complication. The patient was discharged without any problems 2 days after the operation. The patient was regularly followed up via CI mapping. The surgical site, including the left EAC and tympanic membranes, healed well and appear normal. The average hearing threshold using the implant was 35 dB (Figure 1). Compared to previous test when the patient used hearing aids only, the hearing threshold improved. This report was exempted from the research review by the Institutional Review Board of Ajou University School of Medicine (AJIRB-MED-EXP-18-128).

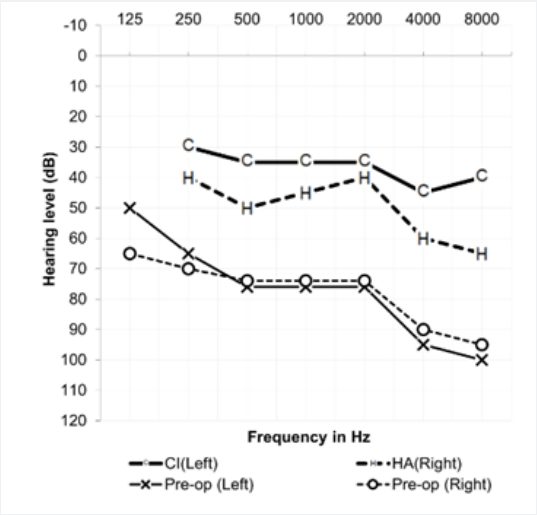

Figure 1: Preoperative pure-tone audiometry and postoperative functional gain testing; the patient had a left-side cochlear implant and used a right-side hearing aid.

Discussion

This is the first report of CI performed in an adolescent with MPS II. We found an earlier case report of CI in a patient with MPS II, but the patient was 4 years of age and no anatomical abnormality was identified [6]. We also suggest, after considering the risks and benefits, that CI is an acceptable option for MPS II patients with SNHL. However, surgical procedures in MPS II adolescents may be confounded by anatomical alterations that progress slowly during the lifespan. Here, we present our experience to share the difficulties that we faced in the surgical procedure. The medical issues of MPS patients are changing; the lifespan is longer than was formerly the case because of early diagnosis, multidisciplinary care, and the introduction of ERT and hematopoietic stem cell transplantation [7,8]. Today, MPS patients live long enough to develop hearing problems. Surgical interventions, including CI, are useful options for MPS patients

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with progressive SNHL. Our present patient was a 13-year-old male adolescent with unexpected anatomical variations. The detailed variations in the middle and inner ears of adolescent or adult MPS patients with SNHL have not been reported. The principal operative problem was venous anomalies in the temporal bone area (Figure 2). Transmastoid emissary and extracranial veins drained the sigmoid sinus. Such anomalies have been reported in patients with CHARGE syndrome [9]. Akira et al. described the performance of CI in a CHARGE syndrome patient [10]. The cited authors used a suprameatal approach featuring cartilage protection; venous anomalies were evident. However, such venous malformations had not been documented in MPS patients when we encountered our patient. Moreover, the paths of the emissary veins varied greatly; it was difficult to obtain comprehensive data even after imaging prior to surgery. Thus, our first operation featured only exploration and vessel ligation.

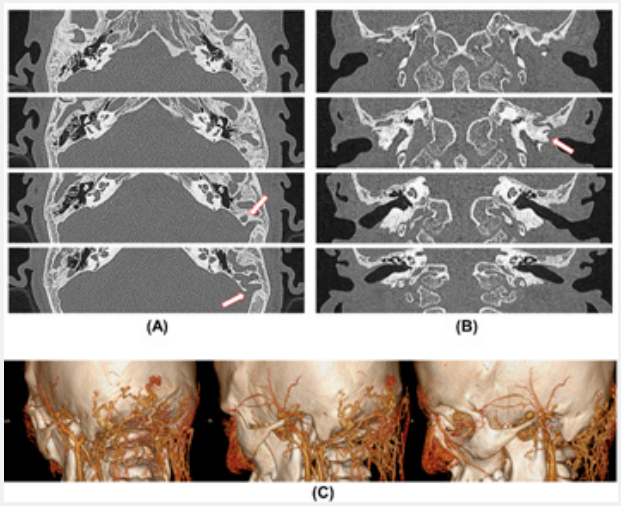

Figure 2: High-resolution temporal bone CT. Axial images (A) and coronal images (B). White arrows indicate emissary veins around the left temporal bone. (C) Temporal bone CT with angiographic 3-dimensional reconstruction. Diffuse, arterial vessel overgrowth is evident around the temporal bone.

The cardiovascular problems of MPS II patients are well known and have been well documented [1,3,5,7,8]. GAG deposition in coronary arteries triggers narrowing or even occlusion and can cause sudden death [11]. Indeed, as GAGs are deposited throughout the entire body, problems with other vessels may be anticipated in MPS patients. To date, only one report has described cerebrovascular changes in an MPS stage IVa patient [12]. Notably, cerebrovascular vessel overgrowth has not been described in MPS II. However, our patient had obvious collateral arteries, as shown in (Figure 2C). Both the emissary veins and collateral artery overgrowth were attributable to vessel narrowing caused by GAG deposition. The current longer life expectancies of MPS patients may change the nature of surgical interventions required by such adolescents or adults, and unexpected vessel overgrowth may compromise surgery. In our experience, CT accompanied by angiographic reconstruction aids preoperative assessment.

Transmastoid emissary veins may bleed profusely, as we found during the first surgery; we thus chose the transmeatal approach to the round window rather than a conventional facial recess approach with mastoidectomy. Like Ganaha et al. and Slavutsky et al., we created an EAC groove to accommodate the electrode array via drilling using an endomeastal approach [10,13]. We used cartilage to protect the electrode array from contact with skin. As reported previously, the EAC was patent and the electrodes functioned well during follow-up; however, long-term observation is essential. In addition, venous vessel overgrowth around the area of the receiver-stimulator may become challenging. Because of the risk of bleeding, the well accepting the receiver-stimulator could not be drilled to an adequate depth. The positions of the screws retaining the device were chosen to avoid vessel injury. After considering the location of the receiver-stimulator and the electrode array in the round window, an array that was 80-90 mm in length was considered appropriate. Array lengths vary by the manufacturer. Thus, all possible device locations must be considered when choosing CIs.

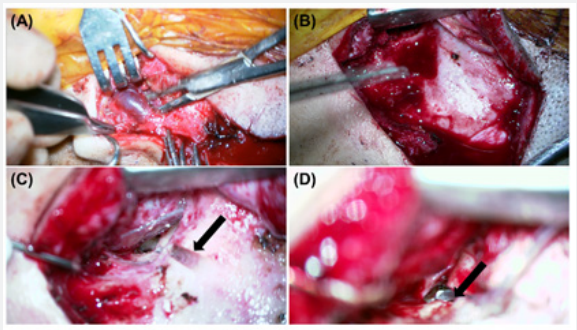

Figure 3: (A) Intraoperative images of the first operation. Emissary veins are apparent between the skin of the scalp and periosteum of the temporal bone. Some vessels derived from the mastoid portion were also identified. (B) All vessels around the operation site were ligated or treated with bone wax. (C) Intraoperative images of the second operation. CI featured the use of a retro auricular trans meatal approach with protection of the cartilage. The skin of the posterior EAC was elevated and the round window exposed (black arrow). (D) The electrodes were inserted with protection of the cartilage (black arrow). The electrodes were then covered with surgical glue and bone wax.

Conclusion

In contrast to CI performed at early age in a prior study, an adolescent MPS II patient may present with unpredicted anatomical abnormalities (e.g., the vessel overgrowth), which may compromise successful performance of the operation. Therefore, a surgeon should carefully evaluate all anatomical abnormalities prior to surgery. The difficulties can be overcome; thus, CI is a feasible treatment option for adolescent MPS II patients with SNHL.

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