CASE REPORT

Cochlear implantation outcome in a child with sensori-neural hearing loss and brain migrational anomaly

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Abstract

Cochlear implantation (CI) is a new treatment for profound sensori-neural hearing loss. Children with syndromes and conditions resulting disabilities are now not routinely precluded for receiving CI. We present an 8-month-old girl with a brain migrational anomaly and profound hearing loss who underwent CI.

Keywords: Brain migrational anomaly, Child, Disabilities, Cochlear implantation, Hearing loss

Introduction

Treatment of children with profound sensori-neural hearing loss (SNHL) has been influenced by diagnostic improvements and technological treatment advances, specifically new cochlear implant prospects recently.[1] Nowadays, multiple handicaped children and children with syndromes and conditions resulting disabilities such as dual sensory loss, brain segmental dysplasia, cerebral palsy, global developmental delay and autistic spectrum disorder are not routinely precluded from receiving a cochlear implant.[2,3] Developing technical facilities and increasing experiences has led more challenging cases are candidates for cochlear implantation (CI). Neuronal migration disorders (NMD) is a diverse group of the human brain malformations that primarily affect the development of the cerebral cortex. Corpus callosum agenesis, lissencephaly, pachygyria, heterotopias, schizencephaly, holoprosencephaly, polymicrogyria and etc., are forms of brain anomalies in NMD.[4] Multiple additional handicaps such as SNHL and motor developmental delay (MDD) may be seen in these patients and they need several special care. Theoretically, CI should be effective in these patients although it did not try formerly.

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Case Report

We present an 8-month-old female infant who was referred to Baqiyatallah cochlear implant center due to bilateral profound SNHL diagnosed by otoacoustic emissions test. In past medical history, she was a product of non-consanguineous marriage without any family history of SNHL and mental or MDD, who was born by cesarean section after an uneventful pregnancy with gestational age of 34 weeks and birth weight of 2250 g. Phototherapy was performed due to mild hyperbilirubinemia during the hospitalization. Regarding to profound SNHL, cochlear implants were offered to her parents. Her evaluation showed MDD based on denver developmental screening test-II.[5] She could not sit without the support and a mild spasticity was detected in her left extremities, her mental and cognitive development was relevant to her age.

Brain magnetic resonance imaging was revealed pachygyria and marked abnormal gyration in right temporoparietal lobe, associated white matter volume loss was present in the obtained coronal T1 images. In Axial T2 section, small microgyria in the frontal lobe supplemented by pachygyria and double band sign was seen [Figure 1]. The entire above are compatible

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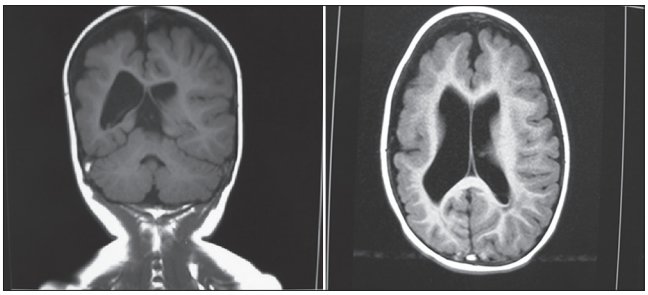

Figure 1: Magnetic resonance imaging of case

with NMD. Serum and urine evaluation for inborn errors of metabolism (such as amino acids chromatography, Lactate and Ammonia) were normal. Thyroid function tests and venous blood gas analysis were in normal ranges. Cytomegalovirus antibody immunoglobulin (Ig) G and IgM was not reactive. Then, she was referred to a rehabilitation center for physical and occupational therapy with the impression of cerebral palsy. The second visit was done after 4 months of rehabilitation, she had acquired sitting skill at 10 months of age, but could not stand independently, her left upper limb and two lower limbs were spastic. Therefore, the rehabilitation therapy was continued until she started walking independently in 2 years of age, although the tonicity of her left limbs was increased and she had left side tiptoe walking.

At this age, she referred for CI and at the post operation follow-up she did not show any CI related complication. Hearing ability was evaluated by the categories of auditory perception (CAP) scale and speech ability was assessed by speech intelligibility ratings (SIR).[6] At 12 months post-operative assessment, the patient, who had 0 score in CAP (no awareness of environmental sounds) at baseline, progressed to category 5 (understanding of common phrases without lip-reading). In addition, the patient SIR score was increased from category 1 (connected speech is unintelligible) pre-operatively to category 4 (connected speech is intelligible to a listener who has little experience of a deaf person's speech) [Tables 1 and 2]. At the last follow-up visit in 24 months after implantation, her neurological and developmental condition was appropriate.

Table 1: Categories of auditory perception scale

Category	Description
0	No awareness of environmental sounds
1	Awareness of environmental sounds
2	Response to speech sounds (e.g. "go")
3	Identification of environmental sounds
4	Discrimination of some speech sounds without lip-reading
5	Understanding of common phrases without lip-reading
6	Understanding of conversation without lip-reading
7	Use of telephone with known listener books

CAP: Categories of auditory perception scale

Table 2: Speech intelligibility ratings

Category	Description
1	Connected speech is unintelligible. Pre-recognizable words in spoken language (primary mode of communication may be manual)
2	Connected speech is unintelligible. Intelligible speech is developing in single words when context and lip-reading cues are available
3	Connected speech is intelligible to a listener who concentrates and lip-reads within a known context
4	Connected speech is intelligible to a listener who has little experience of a deaf person's speech
5	Connected speech is intelligible to all listeners. The child is understood easily in everyday contexts

SIR: Speech intelligibility ratings

Discussion

Almost all patients with a form of NMD are severely neurologically impaired. This neurologic impairment could be involved sensory impairment such as auditory perception disorder however; SNHL of varying severity is not always present.[7,8] Furthermore, regarding to brain structural malformation the patients may be affected by motor neuronal defects. Children with or without genetically based disorders may be affected by SNHL accompanying NMD. Patients with several handicaps are responsible for a small percentage of implanters in a CI schedule because they remain the most challenging cases for which to predict profit from the CI and post-operative rehabilitation.[9] Based on our knowledge, for the first time, we report a child with NMD and profound bilateral SNHL that underwent CI and her auditory perception and speech intelligibility progressed properly.

Conclusion

CI could be an effective option for children with SNHL and neurologic developmental disorders.

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