Cochlear Implant in Deaf and Blind Patient

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Abstract: ***Patients with multiple sensory deficits such as hearing loss and visual impairment (VI) represent a unique problem. The most common syndrome of deaf and blind- Usher syndrome comprises of retinitis pigmentosa and progressive hearing loss. Objective of this study is to evaluate the results and benefits of cochlear implant in deaf blind people.***

Key words: Cochlear Implant, visual impairment, Usher syndrome, CAP Score

1. Introduction

Usher syndrome is an autosomal recessive disorder characterized by a congenital sensorineural hearing deficit of varying severity and a progressive visual loss secondary to a pigmentary retinopathy (retinitis pigmentosa). It was first described by Von Graefe [1] in 3 of 5 siblings in 1858. In 1914, Usher [2] was the first to recognize its hereditary nature in his presentation of 41 families. Usher syndrome is also known as Hallgren syndrome, Usher-Hallgren syndrome, RP (Retinitis Pigmentosa) - dysacusis syndrome, and dystrophia retinae dysacusis syndrome. In the United States, the prevalence of Usher syndrome has been estimated to be 4.4 per 100,000 [3] An estimated 66% of all blind-deaf people in the United States have Usher syndrome [4] 3% to 6% of the congenitally deaf population is considered to have Usher syndrome [5].

2. Case report

A 19 year old, postlingual, male patient presented to civil hospital, Ahmedabad with c/o progressive hearing loss and decreased vision over a period of 5 years.

Visual field examination disclosed peripheral ring scotoma. Electrophysiological examination was performed. Pattern visual evoked response was within normal limits and electroretinogram displayed diminished photopic and scotopic response.His ophthalmoscopy showed lack of pigment in fundus, suggestive of retinitis pigmentosa.

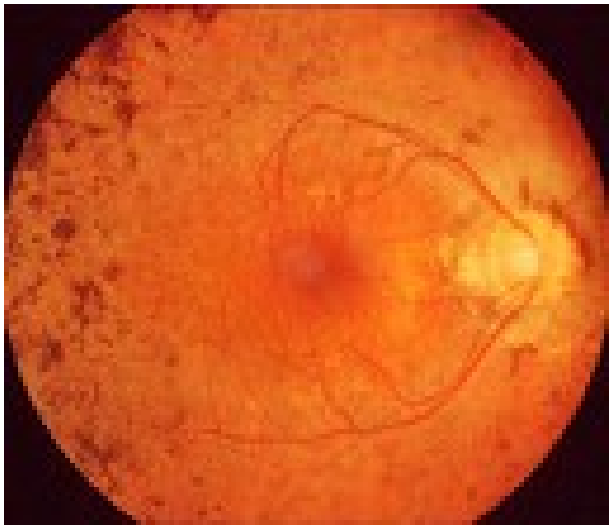

Figure 1: Fundus examination in patient with usher syndrome showing retinitis pigmentosa

Pure tone audiogram (PTA) showed bilateral profound sensorineural hearing loss. Genetic Examination revealed that he had usher syndrome. He was counseled for cochlear implant surgery and further investigated in form of Otoscoustic Emmisions (OAE), Brain Evoked Response Audiometry (BERA), and Auditory Steady State Response (ASSR) which showed bilateral profound sensorineural

hearing loss. CT scan of bony cochlea and MRI brain with 3D reconstruction of membranous labyrinth with 7th, 8th nerve complex, bills bar, brocas area and internal acoustic meatus was normal. A successful cochlear implant was done and the results were evaluated in post operative period.

3. Study Design

Quality of life in a 19 year male deaf and blind patient with usher syndrome type 3 with unilateral cochlear implant was evaluated at our tertiary care center. The mean preoperative and postoperative Pure Tone Audiograms, CAP (Category of auditory perception) score were evaluated

4. Conclusion

Cochlear Implant can play a significant rehabilitative role in patients with severe hearing loss and Visual impairment. Evaluation and rehabilitation sessions are typically more time-consuming and labor-intensive compared with those of normally sighted CI patients. Use of cochlear implant helped him to improve not only his psychological but also his social outlook. The advantage of bionic ear on adding a sense organ to a blind person adds on to compensate his blindness and improving his quality of life.

5. Results

The mean preoperative hearing level (pure-tone average, 0.5-4 kHz) was 100 ± 5 dB hearing loss (HL) and the mean aided hearing level was not in the audible banana spectrum. The postoperative hearing level (30 ± 9 dB HL) and CAP score was 7.

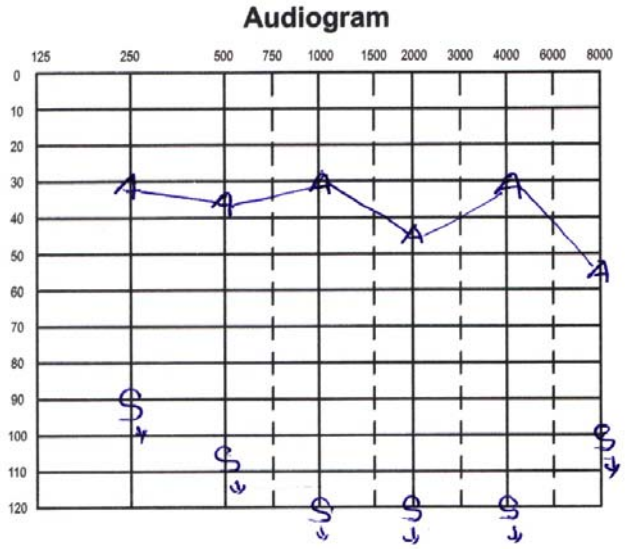

Figure 2: Postoperative hearing level in pure tone audiometry at 1 year is (30 ± 9 dB HL)

The mean preoperative hearing level (pure-tone average, 0.5-4 kHz) was 100 ± 5 dB hearing loss (HL) and the mean aided hearing level was not in the audible banana spectrum.

The postoperative hearing level at the end of one year is 30 ± 9 dB HL.

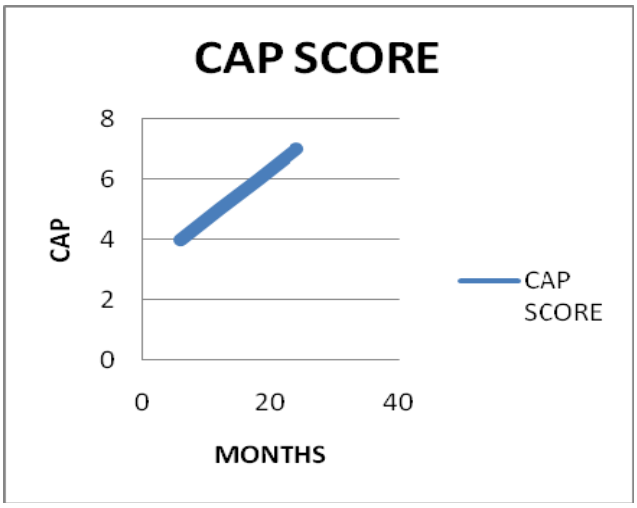

Figure 3: Category of auditory performance (CAP) score

CAP score is a hierarchical rating scale (0-7) which assesses auditory perception skills in a natural context and monitor auditory perception skills over time. This score was assessed over a period of 2 years and the results analyzed. At the end of 2 years CAP Score was 7 (patient can speak on a telephone).

At 2 years:

- Speech identification score is 72-75%
- Speech tracking is approximately from 1 ft.

6. Discussion

As the primary mode of communication for many profoundly hearing-impaired persons is visual (for example, the use of lip-reading, sign language or cued speech).The person with Usher's syndrome may need to learn to communicate through an alternative modality when use of the visual system is restricted. These alternatives are tactile or auditory devices and it may be necessary to learn tactile signing codes. The assessment of the multichannel implant as a sensory substitute is necessary in patients with Usher's syndrome, as well as the assessment of the device as a sensory aid (for use with lip-reading), as the person may lose functional vision by the third or fourth decade of life. Patients' strong motivation for hearing and speaking is important for successful outcome. The performance of patients with profound SNHL and VI is superior to the performance of patients without VI [6] [7] [8]. El-Kashlan et al. [9] postulated that deprivation of one cortical area of sensory input such as vision leads to enhancement of cortical function sub serving another sensory input such as hearing.

7. Conclusion

Cochlear Implant can play a significant rehabilitative role in patients with severe hearing loss and Visual impairment. The success of a cochlear implant depends not just on characteristics of a child prior to implantation, but also on the follow-up intervention and training of the child. Evaluation and rehabilitation sessions are typically more time-consuming and labor-intensive compared with those of

normally sighted CI patients. The follow-up period requires good support from service providers and a strong commitment by the family.

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