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PRACTICALITY OF COCHLEAR IMPLANTATION IN CHILDREN WITH HEARING LOSS AND AUTISTIC SPECTRUM DISORDER IN INDIAN SCENARIO- CASE REPORT.

Audiology

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ABSTRACT

With implementation of new born hearing screening, the age of diagnosis for hearing loss and cochlear implantation age in children has been reduced to 9 months of age by recent FDA criteria. Benefits of early identification and rehabilitation of hearing loss is valuable. But we also need to be aware of the fact that there are chances of missing the diagnosis of other childhood communication disorders like Autism if manifested along with hearing loss, the reasons being the symptoms of autism spectrum disorder as given by Diagnostic and Statistical Manual (DSM V) may not be picked up earlier than two years and also because of diagnostic overshadowing .Some of the symptoms of autism such as deficiency of language, lack of attention, lack of eye contact and clumsiness can be mistaken as symptoms of hearing impairment. This report describes the case study of a 4-year-old female child with hearing loss and autism who was evaluated by a multidisciplinary team of professionals. We call attention to the expected outcome, the need to counsel the caretakers regarding expected outcome, type of therapy and the need for a multidisciplinary team regarding candidacy for CI, continuous support for behavioral management and assessment of outcomes.

KEYWORDS

autism spectrum disorder, congenital hearing loss, cochlear implantation, Rehabilitation

INTRODUCTION

Autism spectrum disorder (ASD) is characterised by persistent impairments in social communication, restricted repetitive and stereotyped patterns of behaviours, interests, or activities [1]. Prevalence of ASD has been reported to be around 2%.[2] Children with ASD have broad range of behavioural characteristics like unusual sensory seeking, repetitive behaviour, stereotyped movements, resistance to changes in the environment, difficulties with social reciprocity and communication. Hitoglou et al (2010) suggested the presence of autism in 5.3% of children with hearing loss. [3]

Congenital hearing loss has a major impact on speech and language development as well as educational achievement. Auditory information received from cochlear implants is far superior to that from hearing aids and hence children with a severe-to-profound hearing loss who use cochlear implants early attain better speech and language skills compared to those using hearing aids [4]. With implementation of new born hearing screening, the most recent guidelines of the Joint Committee on Infant Hearing (JCIH) has changed the standard 1-3-6 guideline of hearing screening by 1 month of age, appropriate evaluation by 3 months of age, and appropriate intervention by 6 months of age to a 1-2-3 guideline[5].

The first FDA approval for CI was in 1990 with the age criterion for candidacy for CI as 24 months of age which was reduced to 18 months in 1998 and to 12 months in 2000 and 9 months of age in 2020[6,7].

Benefits of early identification and rehabilitation of hearing loss is valuable. But we also need to be aware of the fact that there are chances of missing the diagnosis of other childhood communication disorders like Autism if manifested along with hearing loss, the reasons being the symptoms of autism spectrum disorder as given by Diagnostic and Statistical Manual (DSM V) [8] may not be picked up earlier before two years and also because of diagnostic overshadowing .Some of the symptoms of deafness such as deficits in language, lack of attention, lack of eye contact and clumsiness can be mistaken as symptoms of hearing impairment.

Case Report

A 4 year old child born of a non-consanguineous parentage born at full term was brought to the department of Otorhinolaryngology with the complaint of delay in language development and no response to sound stimuli.

Based on the request from ENT surgeon, detailed audiological evaluation was done. Brainstem evoked response audiometry revealed that there was no peak till 90dBnHL. Distortion product otoacoustic emissions were absent bilaterally. Tympanometry revealed bilateral 'A' type tympanogram with absent reflexes bilaterally. Behavioural response audiometry revealed no response to sound till audiometric limits. Hearing aid trial was done and her aided responses were out of speech spectrum.

During her follow up visit, she was recommended for Cochlear implant surgery in the department of ENT.

As part of the routine protocol, the child was evaluated in the Dept. of Developmental Paediatrics, by the Developmental paediatric team, which included a developmental paediatrician, Psychologist and a Speech Pathologist. During the evaluation she was detected to have poor eye contact, stereotypic behaviour, hyperactivity, poor attention and communicated primarily by placing the hands of her parents on the desired objects. She preferred to play on her own. She was then diagnosed to have autism based on the DSM-V criteria. She scored zero on CAP-Revised (Categories of Auditory Performance) [9] and her expressive and receptive language was assessed using The (REELS) Receptive-Expressive Emergent Language Scale-3[10] .

She was found to have an Expressive language age of 0-1 month (Frequent crying and had random vocalizations) and Receptive language age of 0-1 month (no response to sounds) on REELS. The CAP is a hierarchy rating scale which assesses a child's development of speech recognition ability and functional hearing in hearing impaired children. Parents or professionals rate the skills based on their knowledge of the child.

The Receptive-Expressive Emergent Language Scale-3 is a 132-item checklist that uses reports by parents or caregivers to identify major receptive and expressive language problems in infants and toddlers up to 7 years of age.

In view of her hearing impairment and co-existent autism formal developmental assessments could not be completed. However, her social skills and performance skills were assessed using subtests of Griffith's mental developmental scales (GMDS) [11], which revealed that her personal social skill corresponded to 15 months and her performance abilities corresponded to 17 months. The Griffiths Mental Development Scales (GMDS) is used to assess the development of children from birth to 8 years. After completion of the above assessments, she was diagnosed as having bilateral sensorineural hearing loss with ASD (autism spectrum disorder) by a multidisciplinary team consisting of Developmental Pediatrician, Speech Pathologist and Psychologist.

In the department of developmental pediatrics, Parents were counselled regarding the additional disability that the child is having

apart from the hearing loss and was made aware regarding the limited use of cochlear implant in language development of the child. Cochlear implantation was deferred for a few months during which the parents were advised to get the child accustomed to wearing hearing aid and develop tolerance to sound stimulation. she was advised to use widex BV 38 Digital BTE (behind the ear) hearing aid bilaterally and attend auditory verbal therapy. The child was also asked to attend behavioural therapy to improve her attention and cognition through structured play and speech therapy to improve communication abilities. The child was under observation by the team members which also helped in assessing the auditory tolerance to different sound stimuli. Initially there was resistance to wear hearing aids but the hearing aids were put on to the child while working on a fun activity together and slowly made the child accustomed to wearing hearing aids. Six months after the initial diagnosis of autism, once the child started wearing hearing aids without pulling them out, she underwent the cochlear implantation. Switch on was done two weeks later and she was started on auditory verbal therapy. Behaviour modification strategies were continued.

Post-operative evaluation of auditory, language and behavioural skills were done at a three months interval. Her attention had improved, and she had started making good eye contact with her parents, her stereotypic play pattern had reduced. On the GMDS her performance abilities corresponded to 34.5 months and Personal social skill had improved to 19 months.

Her auditory responses were measured using the CAP-Revised 5 (Categories of Auditory Performance,) and her expressive and receptive language was assessed using the (REELS) Receptive-Expressive Emergent Language Scale-3 6 on a three months interval. She had a baseline score of zero on CAP preoperatively which indicates that the child is unaware of environmental sounds. Her scores improved to one during the assessment done 3 months post operatively which indicates that she can detect some environmental sound. However, there was no change in scores when the assessment was done at 6 months, 9 months and 12 months postoperatively.

The child was found to have an Expressive language age of 0-1 month (Frequent crying and had random vocalizations) and Receptive language age- 0-1 month (no response to sounds) on REELS during preoperative assessment. Assessment done 3 months post operatively showed that she had an Expressive language age-1-2 months (cooing and babbling) and Receptive language age-1-2 months (appears to listen to speaker). Nevertheless, her scores did not show any progress when the follow up assessment was done at 6 months, 9 months and 12 months postoperatively.

It was found that even after one year of auditory verbal therapy; the oral language skills of the child did not improve. Hence the decision was made to switch on to total communication instead of auditory verbal therapy

DISCUSSION

If co-existent autism is not identified in a child with hearing impairment, it is likely that symptoms of autism like absence of speech, hyperactivity, poor social interaction and poor communication intent may be mistaken as consequence of hearing impairment. Hence it is necessary for professionals who work with children with hearing loss to suspect ASD when preverbal communication behaviours like poor eye contact, lack of pointing, poor orientation for communication, poor joint attention and delays in language acquisition are afar to what one could expect from children with hearing loss alone. Assessment by a multidisciplinary team consisting of Otorhinolaryngologist, Developmental paediatrician, Audiologist, Psychologist and Speech pathologist will help in the diagnosis and management of such children.

ASD is not a contraindication for CI and should not preclude a deaf child from being considered for CI. But appropriate parental counselling regarding expectations is important. Careful consideration must be given to sensory issues like tolerance of auditory stimulation, since there are chances of total rejection of the auditory processor in these children. A trial period of observation of the child using hearing aids will be useful in assessing the auditory tolerance problem before a decision is made regarding cochlear implantation.

After one year of auditory verbal therapy which emphasizes on aural-oral skill development, since the child did not develop oral language, Parents were counselled that communication using spoken language may not be an achievable long-term goal and may need to adopt a 'total communication approach' and we decided to use "total communication" training. Parent survey that evaluated the benefits of cochlear implantation in children with ASD has reported improvements in awareness of the environment, name recognition, response to verbal requests, and reaction to music and did not show significant improvement in Expressive language and that 50% of the children required special education programmes and 23% required total communication programme [12].

Intervention plans for such children will have to be more individualised addressing specific needs by a multidisciplinary team rather than enrolling them into the usual auditory verbal therapy which a typical hearing-impaired child would undergo after cochlear implantation.

Unlike developed countries, the patients bear the expenses of cochlear implantation in India and many of the lower middle-class families have economic pressure spending almost a year's earnings on such a device and the recurring maintenance cost thereafter. The fact that there is a possibility that the child may not develop speech after cochlear implantation, when there is an associated condition like autism needs to be conveyed during preoperative counselling.

CONCLUSION

Autism spectrum disorders are lifelong conditions. Dual diagnoses of ASD with hearing loss pose additional challenges. In this article, we looked at the challenges of recognizing an autism diagnosis along with hearing impairment; the need for explaining the probability of poor speech and language outcome after cochlear implantation during preoperative counselling and the difference in the intervention needed in these children. We emphasize the need for a multidisciplinary evaluation and management in such children.

Patient Perspective

Parents reported that the cochlear implant device did not help the child in anyways in the speech and language development. The child has a younger sibling and the family finds it difficult managing the recurring expenses of intervention.

Informed consent

The informed consent was obtained from the parents of the child

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