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Auditory and Speech Language Development of a Child with Cochlear Implant with Mondini's Dysplasia: A Case Study

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Abstract

Cochlear implants provide functional hearing to the majority of recipients and have gained wide spread acceptance clinically. The role of the cochlear implants in young children is increasingly being seen as a vital importance for the success of auditory and speech language development. This paper reviews the analysis of the effect of early identification and intervention with unilateral cochlear implant (left ear) on the auditory, speech and language development in a child aged 12 years with Mondin's dysplasia. The study aimed to evaluate and compare auditory and speech language skills pre and post cochlear implant over a period of fifty four weeks. Significant improvement in the studied domains indicate an importance of early identification and role of amplification devices (especially cochlear implants) in the case studied.

Keywords: Mondini's dysplasia; Cochlear implant; Hearing loss

Introduction

Cochlea is named after the Latin word for snail shell because of its coiled snail like shape [1]. The walls are made up of bone with a thin lining of tissue encompassing 3 chambers. The two large chambers include the upper vestibular canal and lower tympanic canal which both contain fluid called perilymph. The two canals are separated by a smaller chamber called the cochlear duct, which is lined with the basilar membrane and filled with fluid called endolymph [2]. At the floor of the cochlear duct is the organ of corti, which is lined with hair cells that act as receptors. Just above the organ is the tectorial membrane. Pressure of vibrations from middle ear stimulates the hair receptors which converts the pressure waves into nerve impulse which is then sent to cerebrum through auditory nerves. Cerebrum then turns signals into sounds [2]. Congenital malformation of these structures is rare anomalies, these occurs due to arrested development at different stages of embryogenesis. A classification first proposed in 1987 by Jackler et al has become widely accepted which divides congenital cochlear anomalies according to the timing of developmental arrest, starting at 3rd week till the end of 7th week of gestation, an insult during each subsequent week results in a distinct inner ear abnormality; at 3rd week- complete labyrinth aplasia (Michel deformity), at 4th week- cochlear aplasia, at 5th week- common cavity to the cochlea and vestibule (cystic cochleovestibular malformation), at 6th week- cochlear hypoplasia (mondini deformity), also known as Mondini malformation or Mondini dysplasia. Mondini dysplasia is an abnormality of the inner ear that is associated with sensory neural hearing loss. The deformity was first described in 1791 by Carlo Mondini after examining the inner ear of a deaf boy [2]. In this anomaly only basal turn of cochlea is developed. There is deficient interscalar septum for distal one and half turn, thus bony cochlea is restricted to 1.5 turns only. The Mondini dysplasia describes a cochlea with incomplete partitioning and a reduced number of turns. A normal cochlea has two and a half turns and a cochlea with mondini dysplasia has one and a half turns. It may occur in one ear (unilateral) or both ears (bilateral) and causes varying degree of Sensory Neural Hearing Loss, most individuals have profound hearing loss [3]. It can also be predisposing to recurrent meningitis because the defect can act as a "port of entry" to the fluid that surrounds the brain and spinal cord (cerebrospinal fluid) [4]. Etiology is disruption in the embryonic development of the inner ear during the 6th week of gestation however the underline cause of Mondini dysplasia in most individuals appears to remain unclear [1]. More recently, a type of mutation called a micro deletion (a tiny loss of genetic material on a chromosome that may span several genes) involving the POU3F4 gene on the X chromosome was detected in some individuals with familial Mondini dysplasia. The condition may be isolated or may occur with other ear malformations and syndromes [5]. It has been associated with thalidomide and rubella embryopathies as well as a number of syndromes such as Klippel Feil syndrome, Pendred syndrome, Di George syndrome [3]. Unfortunately for many cases of isolated mondini dysplasia there is no genetic testing available [6]. It is clear from Hartley's translation that the inner ear anomaly described by the Mondini consists of a cochlea of one-and one-half turns instead of the normal two-and-one-half turns, comprising a normal basal turn and a cystic apex in place of the distal one-and-one-half turns, an enlarged vestibule with normal semicircular canals and an enlarged vestibular aqueduct containing a dilated endolymphatic sac [5]. Arrest at early stages in the spectrum include, cochlear hypoplasia, common cavity and cochlear aplasia. Even earlier arrests result in otocyst and complete labyrinthine aplasia (Michel's anomaly) [7]. Mondini dysplasia is listed as a "rare disease" by the Office of Rare Disease (ORD) of the National Institute of Health (NIH). Treatment options may include surgical repair of the defect to prevent recurrent meningitis; amplification aids for those with residual hearing; and cochlear implantation [6]. The use of cochlear implants to treat

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Table 1: Comparison of auditory skills, speech and language skills pre and post cochlear implantation.

TEST ADMINISTERED	PRE-COCHLEAR IMPLANT	POST-COCHLEAR IMPLANT
Auditory Skill Checklist	Only detection was present, however inadequate.	Achieved auditory skills : 1. Detection 2. Identification 3. Emerging auditory skills: 4. Discrimination 5. Comprehension
Weiss and Curtis	Limited speech and language skills to be assessed.	RLA: 48 months ELA: 36 months
SECS	RLA: 2.0 to 2.11 years ELA: 2.0 to 2.11 years	RLA: 5.0 to 5.11 years ELA: 4.0 to 4.11 years
LPT	Limited speech and language skills to be assessed.	Total language score: 4 to 4.5 years
PAT	90% of misarticulation in child's speech was reported.	50% of misarticulation in child's speech is reported.

patients with mondini dysplasia has been studied variably in recent decade or two. Various results of cochlear implantation in individuals with mondini dysplasia have been reported in the literature and their prognosis varies but some showed greater outcomes in terms of the auditory and speech language development if it is treated early [8].

Need for the Study

Congenital inner ear malformation such as mondini dysplasia acts as an obstruction to the normal auditory development of a child since his/her birth.Need for this present study is to track the outcomes of the early intervention programs in a case having mondini dysplasia.

Aim: Aim of the current study is to assess effect of early identification and intervention on the development of auditory, speech and language skills in a child with Mondini's dysplasia.

Methodology

Subject

Participant of the study was a male child aged 12 years with the history of congenital inner ear malformation diagnosed as Mondini dysplasia. The case reported to Ali Yavar Jung National Institute for the hearing handicapped, NRC, Noida, Uttar Pradesh, with a history of unilateral cochlear implant(left ear) done at the age of 5 years with subsequent sessions of Speech and language therapy.

Educational history

At the time of reporting, the case studied in 4th standard in a regular school set-up with an exposure to Hindi and English language as curriculum subjects.

Test tool

Audiological assessment: auditory development was assessed using an auditory skills checklist developed and validated by University of Cincinnati. The checklist consisted of 35 items to be used by the audiologist or therapist, which relied on a combination of the family's observations of their child's auditory and language skills along with the observations of the managing clinician during therapy/audiology sessions. Based on observed pattern of auditory skill development, items on the auditory skills checklist follows a continuum starting with detection, discrimination, identification and lastly comprehension. These auditory stages are referred to as the comprehensive approach described by Erber (1982). Questions probed a wide range of skill levels, including basic skills such as wearing amplification and showing awareness of environmental sounds. These 4 continuums were evaluated in two groups 'have skills' and 'emerging skills'.

Speech and language evaluation tools: Speech and language development was assessed using four different formal tests which relied on both parent's feedback and observation of the managing clinician. Formal test administered were Weiss and Curtis-Development for language and speech; Scales for Early Communication Skills (SECS) for hearing impaired child given by Jean S.Moog & Ann V. Geer (1975); Photo Articulation Test (PAT) in Hindi, and Linguistic Profile Test (LPT) in Hindi.

Procedure

The case was studied longitudinally over a period of fifty four weeks. Evaluation comprised of parent's feedback and clinician's own observation during speech and language therapy/auditory training sessions. All assessments and evaluations were conducted in sound-treated room set up. To evaluate auditory development, auditory skills checklist was administered and parents were instructed to correctly report about the present auditory development status of their child by answering in yes/no for both sets of questionnaire -'have skills' and 'emerging skills' of the child, while avoiding errors; clinician's observation of the case in the clinical set-up were also considered for scoring. Before the beginning of assessment, the questionnaire was carefully explained to the parents. Set 1 questionnaire comprised of 35 items having questions regarding the auditory skills that are already present in the child i.e. 'have skills'. The questionnaire followed a continuum starting with detection, discrimination, identification and then comprehension. Set 2 questionnaire also comprised of 35 items, having questions regarding the auditory skills that is emerging in the child i.e. 'emerging skills'. This questionnaire followed the same continuum as it followed in Set 1 questionnaire. SCORING: In auditory skills checklist the questions for which parents responded with a 'yes' in both Set 1 questionnaire ('have skills') and Set 2 questionnaire ('emerging skills') was marked and noted by the clinician.

Speech and language evaluation comprised of 4 formal tests procedures. To evaluate the receptive and expressive behaviours of the child, Weiss & Curtis- Development for language and speech was administered with the child. Along with the delayed speech and language child had articulation difficulties as well, to assess the precise discrimination of misarticulation such as substitution, emission, distortion and addition in the child's speech and language, Photo Articulation Test (PAT) was administered in Hindi with the child. For the evaluation of verbal and non-verbal receptive &

expressive language skills (SECS) Scales for Early Communication Skills was administered. To evaluate the semantic and syntactic skills present in the child (LPT) Linguistic profile test was administered in Hindi. Semantics part consists of receptive section, expressive section, semantic discrimination/similarity/contiguity/anomaly, lexical categories, paradigmatic relations, antonyms, synonyms, and homonyms. Syntax part consists of plural forms, tenses, conditional clauses, sentence type, comparatives, and participant construction. All the above formal tests were administered according to the instruction and procedure provided in each scale's manual and with parent's feedback and clinician's observation.

Speech and language formal tests score were calculated as per standard directions from the respective test manuals.

Result and Discussion

The present case study aimed to assess the effects of early identification and intervention on the development of auditory, speech and language skills in a child with the history of congenital inner ear malformation diagnosed as Mondini dysplasia. Before the cochlear implant, case was using strong 'v' cord body level hearing aid. Pre-cochlear implant auditory detection for speech and non-speech sounds were present, however discrimination, identification, comprehension of auditory stimuli were limited. Speech and language development was delayed. Verbally child could comprehend through words and phrases (limited) with the hearing aids- 'ON' & expressed through vocalization and words (limited). Non-verbally child comprehended through signs, gestures and facial expressions and expressed through signs, gestures, pointing and dragging. Percentage of misarticulation was 90%.

Auditory skills checklist evaluation of a post cochlear implant case with the history of Mondini's dysplasia revealed a delayed development of auditory skills in comparison to an age matched typically developing peer. However, the development of auditory detection, discrimination, identification and comprehension were significantly post cochlear implant (Table 1).

Speech and language formal tests results revealed reception at 48 months & expression at 36 months on Weiss and Curtis. On SECS, receptive language age was 5.0 to 5.11 years & expressive language age at 4.0 to 4.11 years. Percentage of misarticulation score was 50% as obtained on Photo Articulation Test. Language receptive score of 4 to 4 & half years and language expressive score of 3 to 3 & half years, total language score of 4 to 4 & half years was obtained on Linguistic Profile Test (Table 1).

The significantly improved outcome of the early intervention program that comprised of regular speech, language and auditory training sessions in the present throws light on the role of early identification along with aural rehabilitation using advanced amplification devices in cases with congenital ear malformations. The study indicates need for easier and mass friendly policies from government bodies and other funding agencies for making the advancement in technologies reach the needy on a wider scale. Also, spreading awareness and counseling the parents of children with congenital ear malformations is highly warranted to achieve the maximum benefits of aids and appliances along with other rehabilitative measures.

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