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The Comprehensive Management and Habilitation Strategy in Late Diagnosed Congenital Hearing Loss with Delayed Speech: A Case Report

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Abstract

Introduction : Hearing impairment in children happened in 6 per 1000 newborn in developing country. The etiology of most of those case is unknown as stated by WHO that 60% of children hearing loss caused by genetics. Aim : This condition could lead to delayed speech in children. Method : A-6-years-old children referred to ENT department with delayed speech and bilateral profound sensorineural hearing loss to get a choclear implant. Result : Her condition suspected to be caused by rubella infection during pregnancy. She was already using hearing aid occasionaly and her mother complain that she still couldn't hear effectively. The comprehensive management of infant with congenital hearing loss could be diagnosed early by applying hearing screening on all newborns at 1 month of age based on EHDI recommendation. This case represents the society low awareness and diagnostic tools limitation of congenital hearing loss prevention in Indonesia. Therefore, we suggest newborn hearing screening on all high-risked babies based on the risk factor stratification. Habilitation strategy for this patient includes auditory-verbal therapy. In order to maximize the benefit, parents are required to give maximum contribution to each therapy session. The therapy compliance becomes the important parameter in outcome optimization. Early diagnosis and intervention is the key to the management of congenital hearing loss. The EHDI programs provide recommended timeline for newborn hearing screenings, diagnostic examinations, and early interventions. The treatment consists of conventional hearing aid and cochlear implantation, combined with speech and language therapy in an appropriate educational environment, to promote development of communication and educational achievements.

Keywords:
Gangguan pendengaran bawaan
Skrinning pendengaran
Strategi habilitasi
Keterlambatan berbicara

Abstrak

Latar Belakang : Gangguan pendengaran pada anak terjadi pada 6 per 1000 bayi baru lahir di negara berkembang. Penyebab sebagian besar kasus tersebut tidak diketahui. WHO menyatakan bahwa 60% gangguan pendengaran anak disebabkan oleh faktor genetik. Tujuan : Kondisi ini bisa mengakibatkan keterlambatan bicara pada anak. Metode : Anak usia 6 tahun dirujuk ke departemen THT dengan keterlambatan bicara dan gangguan pendengaran sensorineural bilateral yang mendalam untuk mendapatkan implan choclear. Hasil : Kondisinya diduga akibat infeksi rubella selama kehamilan. Dia sudah menggunakan alat bantu dengar sesekali dan ibunya mengeluh bahwa dia masih tidak bisa mendengar dengan efektif. Penatalaksanaan komprehensif pada bayi dengan gangguan pendengaran kongenital dapat didiagnosis secara dini dengan menerapkan skrining pendengaran pada semua bayi baru lahir pada usia 1 bulan berdasarkan rekomendasi EHDI.

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Kasus ini merepresentasikan rendahnya kesadaran masyarakat dan keterbatasan alat diagnosis pencegahan gangguan pendengaran bawaan di Indonesia. Oleh karena itu, kami menyarankan skrining pendengaran bayi baru lahir pada semua bayi berisiko tinggi berdasarkan stratifikasi faktor risiko. Strategi habilitasi untuk pasien ini termasuk terapi auditori-verbal. Untuk memaksimalkan manfaatnya, orang tua dituntut memberikan kontribusi yang maksimal pada setiap sesi terapi. Kepatuhan terapi menjadi parameter penting dalam optimalisasi hasil. Diagnosis dan intervensi dini adalah kunci penatalaksanaan gangguan pendengaran kongenital. Program EHDI memberikan jadwal yang direkomendasikan untuk pemeriksaan pendengaran bayi baru lahir, pemeriksaan diagnostik, dan intervensi awal. Perawatan terdiri dari alat bantu dengar konvensional dan implantasi koklea, dikombinasikan dengan terapi wicara dan bahasa dalam lingkungan pendidikan yang sesuai, untuk meningkatkan perkembangan komunikasi dan pencapaian pendidikan.

1. INTRODUCTION

Hearing impairment or hearing loss define as the inability to have equal hearing function compared to someone with normal hearing thresholds of 25 dB or both ears. (Stevenson et al, 2015). Hearing impairment has devastating consequences on life. World Health Organization estimates the number of people with such disability increased eight times from 42 million people in 1985 to 360 million people in 2011, 7.5 million of which are children under 5 years old. (Olusanya et al, 2014). The estimated incidence of permanent congenital hearing impairment in developing countries is 6 per 1000 newborn, that is three times higher than in developed countries.(Patel et al, 2011) Hearing impaired children could have speech and language development disorder that could finally lead to increased risk of physical, social, and emotional abuse. Without prompt management, these children could end-up being adult with embarrassment, loneliness, stigmatization, isolation, depression, occupational, and relationship issues (Sobri et al, 2020) Several countries have implemented the Universal Newborn Hearing Screening (UNHS). Children diagnosed at younger age with congenital hearing impairment have better quality of life in later life. (Sobri et al, 2018) Indonesia has not implemented any newborn hearing screening in its health policy thus making hearing impairment a national burden. Late-diagnosed children have far less language age than other children. In this review, a case of sensorineural deafness in 6-year-old child is presented with further discussion on current management.

Case Report

A 6-y e a r -old girl was referred to ENT Department of Cipto Mangunkusumo Hospital with delayed speech and bilateral profound sensorineural hearing loss. The hearing loss is suspected to be caused by congenital rubella due to the presented of fever and skin rash on her 28-year-old mother 2 weeks before she got positive result on her pregnancy. There was no complications during pregnancy and delivery. She was born healthily, without report of asphyxia, jaundice, nor cyanosis. The patient and her mother was discharge after three days in hospital. Her growth and motor milestones had always been normal. There was no family history of hearing loss and no other significant medical history. At the age of 20 months, her mother noticed that she was slow on speech. She couldn't say any word and only produce gibberish sound. Her mother said she hardly showed responses to any sound, but getting startled because of a very loud sound. Then she was brought to a pediatrician, and she was referred to ENT, cardiologist, neurologist, and ophthalmologist because she was suspected from congenital rubella syndromes. The assessments from cardiologist, neurologist, and ophthalmologist shown normal, while she was found suffering from bilateral profound sensorineural hearing loss from the hearing assessment. At the age of 2 years and 6 months old, she started using hearing aid, but she often took it off due to the uncomfortability. Her mother said, even with hearing aid, she still couldn't hear effectively. She could only understand what people said with the help of lip reading. Her speech development start to improve, and she started to spoke simple words such as calling her parents and vocalizing what she want, but it was still difficult for her to make a full sentence. She started getting speech therapy since 5 years old and using the hearing aid full time. Despite hearing and speech difficulties, she was enrolled to kindergarten for normal children and she could play well with her friends. Her parents said now she improves more and starts to learning alphabet and reading, but she only responds to loud voice and still depends on lip reading. The otoscopic examination revealed a normal tympanic membrane in both ears. Tympanometry results, shown in Figure 1, revealed a normal middle ear functioning, characterized by type A tympanogram. The otoacoustic emmision (OAE) test result is "refer" for both ears so she need further assessment which is BERA test.. For a concrete evaluation of the auditory function, she proceeded to get BERA (brainstem evoked response audiometry) examination, whose results showed in Figure 2. The absence of V waves was observed on both ears even until 90 dB intensity, these changes being suggestive for a profound sensorineural hearing loss. The second test performed for

confirming the initial diagnosis, was an auditory steady-state response (ASSR) (Figure 3). The examination result sustained the diagnosis of bilateral profound sensorineural hearing loss.

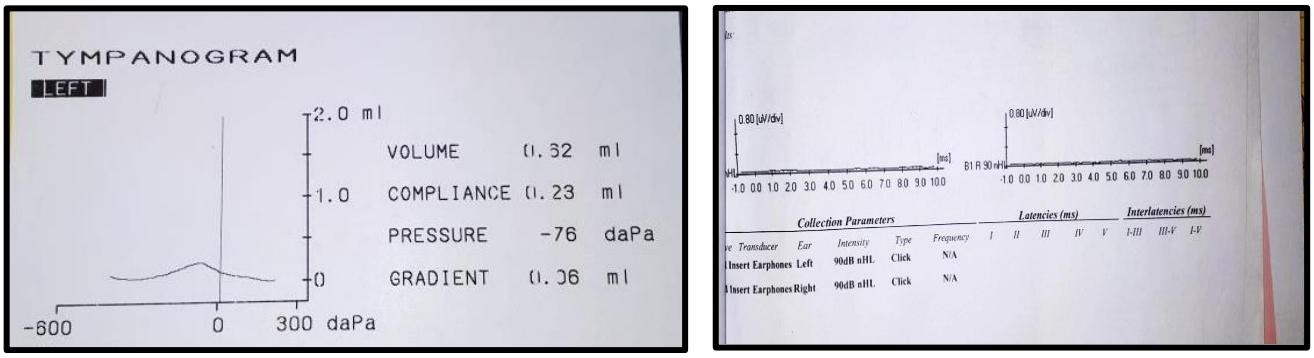

Figure 1. Tympanometry results – bilateral type A tympanography

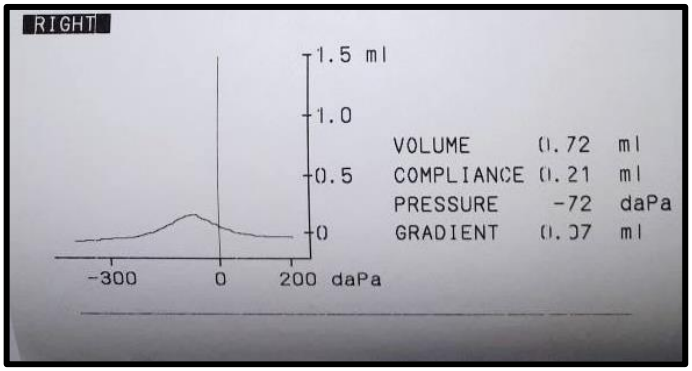

Figure 2. BERA results - absence of V wave until 90 dB intensity

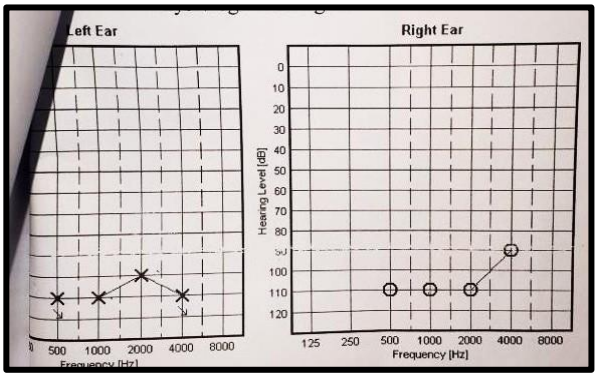

Figure 3. ASSR results. Bilateral profound sensorineural hearing loss

Figure 4. MRI Mastoid

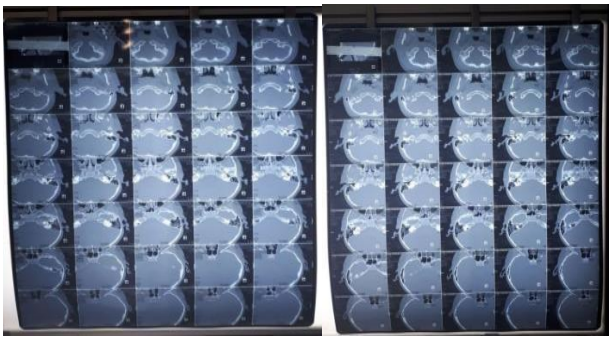

Figure 5. CT Scan Mastoid

She get MRI and CT Scan mastoid investigation before surgery. The MRI reports (Figure 4) symmetrical hyperintensity on white matter of parietooccipital lobes bilateral, and normal structure of cochlea, vestibulum and semicircular canals, while the CT Scan reports (Figure 5) shown normal structure of bilateral middle ear and inner ear. Those results suggesting that the patient could undergo the cochlear implantation.

2. METHOD

Participants were recruited from Cipto Mangunkusumo Hospital. It was clearly stated that the researchers conducting this research were interested in completing "a Comprehensive management and habilitation strategy in late diagnosed congenital hearing loss with delayed speech". To be included in the study, individual had to be congenital.

The study, which was approved by the Universitas Indonesia review Board, had two parts that were counter-balanced across participants. One part focused on self-report hearing loss and second part included screening the partipicants. The goal of hearing screening was not to identify individuals with even a minimal hearing loss, who most likely would not report communication difficulties, seek professional help, or comply with treatment recommendations.

RESULST AND DISCUSSION

The Interconnection of Hearing and Speech Physiology

Hearing process starts from sound received by auricles, travel along the ear canal, vibrate the tympanic membrane, and amplified by the ossicles. Vibration of the ossicles is then transmitted to the cochlea which convert the mechanical wave into electrical signal. This signal is then transmitted to the brain through the cochlear nerve fibers, and perceived as sound. This, other than the visual input, is the first sensory input that infants could get from their environment. Both auditory and visual input influence the ability of an infant to learn how to speak. To speak about something seen, the brain transfers the visual information from the primary visual cortex to angular gyrus parietal-temporal- occipital association cortex which integrates inputs such as sight, sound, and touch. To speak about something heard, the brain transfers the auditory information from the primary auditory cortex to the angular gyrus. This information is transferred to the Wernicke's area, where the choice and sequence of words to be spoken are formulated. The language command is then transmitted to Broca's area where translation of the message into a programmed sound pattern occurs. Signal from the Broca's area is conveyed to the primary motor cortex that activates muscle responsible for the production of word.

Newborn Hearing Screening

Newborn hearing screening leads to earlier detection and intervention in patient with congenital hearing impairment. Therefore, language acquisition and educational achievement in affected patients can be improved significantly through early intervention. (Porter et al, 2009) Screening newborn in earlier age is crucial because caregivers and clinicians are not able to detect hearing impairment until they show delayed speech and language milestones. Two electrophysiologic techniques for newborn hearing screening are automated auditory brainstem response (AABR) and otoacoustic emission (OAE). Both can detect hearing impairment of >35 decibels (dB) and are reliable in infants 3 months of age. They evaluate the cochlea and the peripheral auditory system, but are not able to assess the central auditory system. The AABR has an advantage of being able to detect auditory neuropathy (AN) hearing impairment. (Lin H-C et al, 2005) Universal newborn hearing screening (UNHS) is the current and preferred method used to screen hearing impairment in newborn, the goals are to get early recognition and treatment and maximizing linguistic competence. (Holster et al, 2009) The guideline of UNHS includce Early Hearing Detection and Intervention (EHDI) 1-3-6 recommendation that every newborn has to be screened before they reach one months of age, hearing assessment of all infants who fail the screening test by three months of age, and intervention for those patients with significant hearing impairment by six months of age. (Probst et al, 2012) The two types of screening protocols are single- or two- stage UNHS. The single protocol conducts one screening test, either OAE or AABR, whereas two- stage screening protocol has two steps of screening, and only patients who fail both screens are referred for audiologic assessment. The two-stage protocol is preferred as it can reduces the rate of false-positive test and referral rate. (Dworsack-Dodgen et al, 2012) In this case, the patient didn't get hearing screening which resulting in late detection and intervention. The hearing impairment was assessed by the age of 20 months after the mother complained about her daughter's delayed speech. The hearing screening policy is needed since there is no national program for hearing screening in Indonesia.

Pediatric hearing screening with behavioral audiometric methods

Behavioral audiometric methods conduct to test subjective auditory response in pediatric audiology. These methods are also used in rehabilitative measures, such as hearing-aid fitting. These methods are classified into reflex audiometry, response audiometry, and pediatric speech audiometry. Reflex audiometry is observing the response of infants to auditory stimuli. Those reflexes including sucking responses, motor responses such as Moro reflex, and changes in respiratory pattern. And then in response audiometry, by about 5 months of age, acoustic stimuli can elicit typical response pattern in normal infants. The infant will turn its head toward a sound source. Initially, this response present in the horizontal plane and later occurs in the vertical plane too. Response audiometry can be conducts with distraction test or visual reinforcement audiometry (VRA). (Chen MM et al, 2016)

The next methods is play audiometry. This test can be conducted to 1-2 years old children, because they are already develop their capability to incorporate the tasks and response od pure-tone audiometry into a play setting. And the last methods is pediatric speech audiometry. This kind of test can be used to 2 years old children or older. They will get the instruction verbally to select a certain toys or pictures. This technique can be used to screen for speech recognition problems by telling the child, at increasing distances, to objects displayed in pictures. After 3-4 years of age, we can use audiometric speech tests specially designed for children- e.g., the Pediatric Speech Intelligibility (PSI) test. (Sobri et al, 2020) In this patient, her mother came to the physician for hearing assessment at the age of 20 months, so the simple screening we can conduct at that moment was with play audiometry. Since the patient showed no response to auditory stimuli this behavioral audiometric methods didn't enough to diagnosed the patient. However, the behavioral audiometric methods can be used for hearing aids or cochlear implant fitting.

Auditory Examination Analysis

The otoacoustic emission test (OAE) becomes the first line of auditory functional screening in infant and children. The aim of the test was to examine the cochlea function based on the electrophysiologic mechanism. The OAE result on both ears indicates some dysfunction on both ears, with the suggestion test to be referred to ENT department. (Sobri et al, 2020) Based on the examination preference to rule out the possibility of conduction hearing loss in patient, tympanometry examination was performed as the initial modality to examine the condition of middle ear. The abnormality result indicates some blockage or obstruction in that structure which will result in negative pressure on the middle ear. There were four kinds of abnormal result in this modality. Type AS indicates the hearing bone rigidity with normal ear pressure and decreased compliance, usually seen in otosclerosis patient. Type AD indicates the hearing bone discontinuity with normal ear pressure and increased compliance. Type B indicates the reduced eardrum mobility, with flat curve of pressure and compliance, usually due to the fluid presence in middle ear or tympanic membrane perforation. Type C indicates the dysfunction of Eustachius tube with negative pressure and normal compliance, usually due to obstruction inside the tube. In this patient, the result of tympanometry of this patient is Type A, with normal pressure and compliance in both ears. The CT scan of the mastoid becomes the alternative modalities to rule out the possibilities of conduction hearing loss. In this patient, the CT scan result indicates the hearing bone and mastoid of both ears within normal condition. Based on both examinations, there is slight possibility of conduction hearing loss in this patient. On the other hand, brainstem evoked response audiometry (BERA) become the main preference modalities to rule out the possibility of sensorineural hearing loss. The test was applied to examine the integrity of auditory system based on the electrophysiology principle. The evoked potential triggered by tone-burst or insert probe was analyzed into five levels of positive deflection wave based on the morphology, latency period, and wave amplitude. The main concern in BERA test was the latency period between the stimulus and evoked potential There are three different latency period, which consist of absolute latency period, inter-wave latency, and interaural latency. The presence of latency period elongation will indicate conduction impairment of the ear. On this examination, the V wave of the patient could not be detected till 90 dB. This result indicates the possibility of sensorineural hearing loss in the patient. The auditory steady-state response test (ASSR) is an auditory evoked potential that can be used to analyze hearing sensitivity in individuals with various degrees of sensorineural hearing loss. (Walker et al, 2013) The usage of ASSR is more preferable than audiometry brain stem response (ABR) due to frequency- specific information and wider range of hearing threshold levels. In this patient, the result of ASSR indicates the very severe degree of sensorineural hearing loss in both ears with measured wave intensity at 107.5 dB. The free field test has function in identifying the ability in stimulus response based on some activity test and evaluated by neometer or *Viena tone*. The relation between ASSR and FFT is to measure the amount of functional gain at each specific frequency based on the voluntary behavioral procedures. The FFT aided result on this patient indicated the conventional hearing aids could not be used as single therapy in this patient. It was based on the inadequacy on achieving desirable minimal sound intensity. The MRI scan of the mastoid become the alternative modalities to rule out the possibilities of sensorineural hearing loss. In this patient, the MRI scan result indicates the symmetrical white matter hyperintensity in white mater of the parietooccipital bilateral lobe. The facial and vestibulocochlear nerve of both ears was intact without any narrowing or nerve compression. In addition, the morphology of the cochlea, vestibulum, and bilateral semicircular canals were within normal condition.

The Benefit of Early Interventions in Children with Congenital Hearing Loss

Joint Committee on Infant Hearing promotes universal screening of early hearing detection and intervention (EHDI) for all newborns. EHDI aims to (1) have all infants undergo a newborn hearing screening prior to 1 month of age; (2) those who do not pass the initial screening have a comprehensive audiologic evaluation by 3 months of age; and (3) interventions be implemented to those with impaired hearing by 6 months of age. (Sobri et al, 2018) Early intervention is one of the key to the management of congenital hearing loss. Treatment primarily consists of amplification using conventional hearing aids and cochlear implantation. Amplification is provided to give an opportunity to a child with impaired hearing an access to the auditory environment –and in particular speech, as much as feasible. (Sininger et al, 2020) Appropriate amplified auditory input maximizes the opportunities for the children with impaired hearing to develop age-appropriate receptive and expressive oral communication, language development, and psychosocial skills. (White KR et al, 2004) Thus, the use of hearing aids also minimize the negative impacts on communication development and academic achievement. (Khairi et al, 2015) The challenging fact about this case is that the patient was presented to the hospital at the age of 16 months to get her hearing checked. Based on EHDI timelines, this patient was supposed to get her intervention already at that age. The patient is diagnosed bilateral profound hearing loss from her auditory brainstem response (ABR) examination which showed no response even at 90 dB for both ears. She was diagnosed the age of 16 months. Late diagnosis in this case resulting in late intervention was done.

Hearing Aids

Amplification system is considered for any degree of hearing impairment that can possibly affect normal developmental processes. Therefore, mild or unilateral hearing loss is also considered for hearing aid usage. (Khairi et al, 2015) Bilateral hearing aids are recommended for children with bilateral hearing impairment because they enhance binaural hearing, and improve auditory localization and speech understanding in noise. Age of the patient at amplification fitting affects speech perception and speech production. A study conducted at Boys Town National Hospital compared language competency between children enrolled in intervention program prior to 6 months of age to children enrolled in the program after 6 months of age, showed that the children identified before 6 months of age had a slight advantage over the later ones. The differences between the two groups became larger over time so that by the time these children were almost 5-years old, those enrolled in intervention program prior 6 months of age were achieving the normal range of hearing. (Tharpe et al, 2018)

The effectiveness of hearing aid usage in children is affected by maternal education level, chronologic age, and degree of hearing loss. There is a difference of hearing aid usage in children whom mothers are college graduated with those with mother high-school (or lower) graduated. Focused counseling on hearing aid adherence is therefore needed, especially for families with lower education levels. (Bond et al, 2009) Children with milder hearing loss are less likely to wear their hearing aids in public situations compared with children with more severe impaired, even though mild hearing loss is also significantly associated with poorer academic performance. (Niparko et al, 2010) American Academy of Audiology recommended behind-the-ear (BTE) style hearing aids as preferred choice for pediatric patients. This style is recommended for young children because there is less swallowing risks compared with in-the- ear (ITE) hearing aids as the outer ear size in children continue to grow well into puberty. (Sampaio et al, 2011) Pediatric Working Group suggests that audiologic appointments should be made every 3 months during the first 2 years of hearing aid usage to monitor auditory status and hearing aid fitting as ear canal grows rapidly during early childhood. (Peters et al, 2016) This timeline can be adjusted if there is an increased risk for progressive hearing loss. (Reeder et al, 2015) In this case, the patient was using hearing aids for the first time at the age of 2.5 years old. Even though the patient was diagnosed with profound hearing impairment (i.e. a potential candidate for cochlear implantation), she still should receive a hearing aid trial prior to implantation to determine whether there is a sufficient benefit from hearing aid usage. The absent of response checked by ABR does not exclude a child from hearing aid candidacy as residual hearing may exist at intensity levels greater than standard ARB capable to elicit. (Lieu et al, 2010) The patient started to experience the benefit of hearing aid usage when she attended speech therapy at the age of 5 years old. Beforehand, she often removed the hearing aid due to discomfort causing the benefit of the hearing aid was not achieved maximally. In 2016, the patient was checked auditory steady-state response (ASSR) for both ears and the result was she has bilateral profound SNHL 107.5 dB. The patient was then scheduled to have cochlear implantation on March 2018.

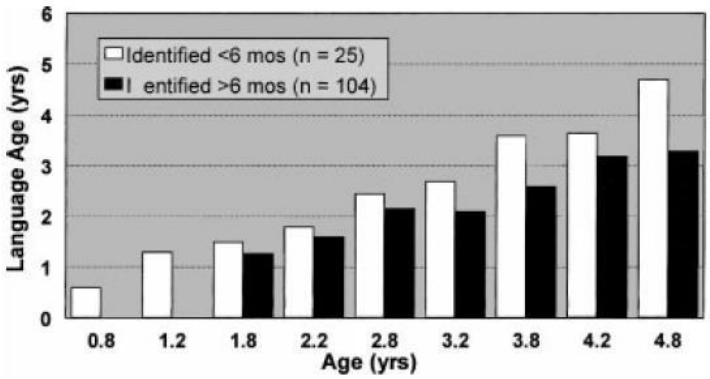

Figure 5. Study of Boys Town National Research Hospital compared language competency of children enrolled earlier in intervention program versus children enrolled later.

Cochlear Implants

Children with severe or profound hearing impairment may not achieve sufficient levels of aided audibility and speech discrimination ability with hearing aids. (Egilmez et al, 2015) Therefore, those children should be referred for a cochlear implant evaluation in order to support the development of auditory skills and speech understanding as cochlear implantation is proven clinically effective in children with severe to profound bilateral hearing loss. Niparko et al. conducted a study of spoken language outcomes in severe to profound SNHL children with cochlear implantation and found that the use of cochlear implants was associated with improved spoken language development compared with their estimated scores from their pre-implantation ones. The use of cochlear implants have also been demonstrated cost-effective. Cochlear implantation can improve speech and

sound perception, promote development of verbal language and auditory skills and educational development, and also suppress tinnitus in some patients. The benefit of cochlear implantation in children with unilateral SNHL is uncertain to date as the result of studies is still mixed

However, study by Reeder et al. showed that children with unilateral hearing impairment have significant differences of speech perception and localization compared with their peers with normal hearing. Children with unilateral hearing impairment have also been reported to have worse language skills than their normal-hearing siblings.The cochlear implant directly stimulates the first order neurons of the auditory pathway, i.e. the spiral ganglion cells. The device is surgically placed into the cochlea and converts sound to an electrical signal which then conducted via electrodes to the spiral ganglion cells in the cochlea. FDA has approved the use of cochlear implants in children beginning at 12 months of age. However, there continues to be a trend toward improved auditory rehabilitation and language development with earlier cochlear implantation. Colletti et al. analyzed the long-term outcomes of cochlear implants in pediatric patients and found that patients implanted before months of age have greater receptive language growth than those implanted at or after 12 months of age. FDA has great influence on cochlear implantation candidacy as it has pursued many clinical trials about this issue. Patients with severe- to-profound bilateral SNHL and some pathologies limited to the cochlea are potential candidates for cochlear implantation. The other criteria are optimal age should be between 12 months and 5 years (but younger ages can also be candidate), lack of improvement in hearing and speech benefits from hearing aids after a 3 months of use, the family of the patient will support the surgery and postoperative rehabilitation, the family and the patients are available for rehabilitation and education, and there are no contraindications for surgery.

The patient in this case met the criteria for the cochlear implantation i.e. bilateral profound SNHL. According to her mastoid CT scan and mastoid MRI result, there is no cranial nerve VIII or brain stem lesions. between ears in order to achieve maximal binaural hearing benefits Map is a program created to set the threshold levels (T -levels) and comfort levels (M or C levels) for patient with cochlear implant. The minimal amount of electrical stimulation required for the auditory system to perceive sound is termed T- levels. The upper limit of electrical stimulation judged to be most loud but is also comfortable is determined by the M or C-levels. Regular scheduled visit should be taken by patient to get maximum benefit. In this case, the patient is scheduled to be checked once every three months for the first year of cochlear implant usage, and once a year for the following year. MRI is superior for detecting cochlear nerve dysplasia and other intracranial pathologies that may impact the implantation of cochlea. However, the best imaging for pediatric cochlear implant candidates is still unclear because majority of anatomic anomalies that would affect cochlear implantation can be seen on either CT scan or MRI.

Table 1. Indication of cochlear implantation based on audiometric candidacy

Indication of the Cochlear Implantation	
12-24	**>2 y**
Profound SNHL	Severe to profound SNHL
PTA > 90 dB	PTA > 70 dB

Contraindications to cochlear implantation include hearing loss due to cranial nerve VIII or brain stem lesions. The other absolute contraindications are severe mental disease, severe mental retardation causing inability to cooperate with speech trainings, and acute or chronic otitis media and mastoiditis without eradication of the disease. The relative contraindications are certain Children with bilateral hearing impairment who receive a cochlear implant in one ear should be fit with a hearing aid in the other ear to require stimulation in both ears. This aims to encourage auditory development and to perform binaural hearing functions. Binaural hearing improves speech perception in noise and sound localization. Thus, binaural hearing also improves functional performance in real life and perceived quality of life. The hearing aid should be optimized with the cochlear implant by balancing loudness of sounds medical conditions like pulmonary, cardiac, and hematologic conditions, uncontrolled epilepsy, and not being available for rehabilitation. (Colletti et al, 2009) Cochlear implantation has a low rate of complications. (Chen et al, 2009) The most common complication of cochlear implantation is infection with the most common pathogen is *Staphylococcus aureus*. Major complications are rare, e.g. facial nerve injury/damage 0.39%, perilymph gusher/cerebrospinal fluid fistula 0.25%, and meningitis 0.11%. The patient in this case underwent cochlear implantation on her right ear in March 17th, 2018. On polyclinic check-up 5 days after surgery, there is no sign of local or systemic infections. Patient is scheduled to have his device switch-on on April 3rd. The parents of the patient have also prepared special hearing environment for the patient by moving her to a special school for impaired hearing children.

60% of childhood hearing loss is preventable

Hearing loss in children has many causes and the exact cause may be impossible to determined. The causes are classified into hereditary genetic causes and acquired causes. Those genetic causes are responsible for nearly 40% of childhood hearing loss. 90% of these hearing impairment are inherited as an autosomal-recessive

trait, so it is more frequent in children born of a consanguineous marriage. Rarely, multiple gene loci or chromosome are affected resulting hearing disorder accompanied by other symptoms. That means the hearing loss is one feature of congenital syndrome. Approximately, 30% of congenital hearing loss are being syndromic, and the remaining 70% being non-syndromic. Hearing loss also can be acquired before birth (intrauterine, prenatally), during birth (perinatally) or after birth (postnatally). Over 30% of childhood hearing loss is caused by infectons, such as rubella, cytomegalovirus, mumps, meningitis measles and chronic ear infections. Meningitis and rubella account for over 19% of childhood hearing loss. Most of those infectio ns can be prevented by immunization and good hygiene. Ear infections also can be prevented through good ear care and general hygiene. Complications at birth, such as asphyxia, low birth-weight, prematurity, and jaundice are responsible for 17% of hearing loss in children. Such conditions can be prevented through safe maternal and child health practices. Use of ototoxic medicines in pregnant women and children is responsible for 4% of childhood hearing loss. (Krug et al, 2016) The risk factor present in the patient is probably rubella infection in early pregnancy. The mother was suspected for rubella infection and already completed her treatment before she noticed her pregnancy. Infection with rubella virus characterized by non-specific signs and symptoms including a transient erythematous pruritic rash, low-grade fever, arthralgia, and lymphadenopathy. This viral infection is highly contagious and transmitted by droplets. (Arumugam et al, 2015) Hearing impairment, either alone or in combination with other defects, is one of the most common abnormality associated with congenital rubella. Congenital hearing impairment may occur after maternal infection up to the nineteenth week of pregnancy. There are several studies which indicated that profound hearing impairment mostly happened when rubella infection occurs in the second month of gestation. Sensorineural hearing loss is the most common defect resulting from intrauterin infection of rubella, and usually it is bilateral. The fetus is being infected transplacentally as a result of maternal viremia after primary infection in pregnancy. Thus, to prevent the occurence of congenital rubella syndrome, WHO has recommended that rubella vaccionation as a part od national immunization program for all children at the age of 12-18 months. Furthermore, for adult protection, ideally all of non-pregnant women with child-bearing age should receive the rubella vaccine too. (Cohen et al, 2014)

Auditory-verbal Therapy

Auditory-verbal therapy (AVT) is a method of encouraging deaf children to listen (auditory) and to speak (verbal) using their hearing capability with the use of hearing aids without depending on lip reading and sign language. Meta analyses from 1993-2015 concluded that AVT has positive impact on developing speech and language skills in children with hearing impairment. (Rahman et al, 2002) Hearing impaired infants do not have the same language environments as their peers due to the lack of auditory input. There is evidence that the longer the brain develops without auditory stimulation, the larger the sensory deprivation, and this can lead to the absence of sensory stimulation to the brain. Some infants maximized visual stimulation provided by the environment and this explain why children using lip reading and gestures for their language processing. (Kaipa et al, 2016) At age 4 years, brain plasticity decreases and the ability to learn spoken language is harder. (Lim et al, 2005) The primary goals of AVT is to develop the same receptive and expressive language skills in children with hearing impairment as their peers with normal hearing. This habilitation strategy needs active involvement of the parent and continuous repetition. (Yoshinaga-Itano et al, 1998) In AVT, children are first though learning to listen sounds, in which parent pronounce words that are used to represent objects for instance “moo” for a cow. “broom broom” for a car, etc. After that, the session could go for more challenging tasks. It is important that parent complete the Ling six sound test three times a day to ensure the hearing aid and cochlear implant is working properly. Ling 6 sound test is a test of listening that is done periodically throughout the day where the child need to respond to 6 sounds (ah, oo, ee, sh, s, and m). The child completes an action or imitates the sound when heard. These sounds represent the sounds across the frequency range for speech. (Eriks-Brophy et al, 2004) In this case, the patient is going to follow an AVT course after switching on the cochlear implant, and after it works efficiently for the patient. The purpose of AVT in this case is to make the patient less dependent to lip reading and sign language. Since this patient is late-diagnosed and late-treated, we cannot yet to determine the success of AVT. Follow up is needed to see whether the AVT is efficient in helping the patient to reach her appropriate language age.

CONCLUSION

Early diagnosis and intervention is the key to the treatment consists of conventional hearing aid and cochlear implantation, combined with speech and language therapy in an appropriate educational environment, to promote development of communication and educational achievement.

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