Characteristics of Infants and Young Children With Sensorineural Hearing Loss in Dr. Soetomo Hospital

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Research Report

Characteristics of infants and young children with sensorineural hearing loss in Dr. Soetomo Hospital

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ABSTRACT

Background: Hearing loss is one of the congenital abnormalities frequently found in children, which is followed by delayed speech and language development. The majority of cases have unknown causes of hearing loss resulting in late diagnosis. Newborn Hearing Screening Program (NHSP) recommended Otoacoustic Emissions (OAE) and Brainstem Evoked Response Audiometry (BERA) as detection of hearing loss in infants and children. **Objective:** To obtain the prevalence and description of sensorineural hearing loss in infants and children. **Method:** A retrospective descriptive study of infants underwent OAE and BERA between 2011-2013 at Dr Soetomo Hospital. The degree of hearing loss was according to the International Standard Organization (ISO). **Result:** A total number of 552 infant and children were examined, and 377 (68%) were detected with sensorineural hearing loss (SNHL). This group of SNHL consisted of 199 males (52.79%) and 178 females (47.21%). The largest age group was 12 to 36 months, revealed 237 patients (62.86%) with SNHL. The majority degree of hearing loss was profound hearing loss in 329 patients (87.27%). The risk factors of SNHL mostly were not found, in 310 patients (82.23%). The majority number of SNHL was bilateral, in 357 patients (94.69%). **Conclusion:** SNHL was found in majority of infant and children in the Audiology Clinic of Dr.Soetomo Hospital. The hearing loss found were mostly profound and bilateral, with unknown risk factors, which might contribute to speech and language developmental delay. This is relevance with the Universal NHSP recommendation that early detection should be implemented to all newborn.

Keywords: hearing loss, OAE, BERA, infant, children

ABSTRAK

Latar belakang: *Gangguan pendengaran adalah salah satu kelainan kongenital yang sering ditemukan, dan berpengaruh pada perkembangan bicara dan bahasa anak. Sebagian besar gangguan pendengaran tidak jelas ada faktor risikonya, sehingga tidak segera terdeteksi. Bila tidak dilakukan deteksi dini, akan menyebabkan keterlambatan diagnosis dan intervensi. Telah direkomendasikan oleh Newborn Hearing Screening Program (NHSP) pemeriksaan Otoacoustic Emissions (OAE) dan Brainstem Evoked Rresponse Audiometry (BERA) sebagai alat deteksi dini gangguan pendengaran pada bayi dan anak.* ***Tujuan:*** *Mendapatkan prevalensi dan deskripsi gangguan pendengaran sensorineural pada bayi dan anak.* ***Metode:*** *Penelitian deskriptif retrospektif dengan mengumpulkan data subjek periode 2011-2013 di Rumah Sakit Dr.Soetomo. Pemeriksaan OAE menggunakan Distortion Product Otoacoustic Emissions. Pemeriksaan BERA berdasarkan International Standard Organization (ISO).* ***Hasil:*** *Terdapat sebanyak 377 pasien (68%) dengan gangguan pendengaran sensorineural dari total 552 bayi dan anak. Pada kelompok umur 12 sampai 36 bulan didapati gangguan pendengaran sensorineural tertinggi sebanyak 237 (62,86%) pasien. Sebagian besar pasien laki-laki sebanyak 199 (52,79%). Mayoritas pasien mengalami gangguan pendengaraan derajat sangat berat sebanyak 329 (87,287%) dari total 377 penderita. Mayoritas faktor risiko dari gangguan pendengaran yang tidak diketahui sebanyak 310 kasus (82,23%), dan mayoritas penderita mengalami gangguan pendengaran sensorineural bilateral sebanyak 357 (94,69%).* ***Kesimpulan:*** *gangguan pendengaran sensorineural ditemukan terbanyak pada bayi dan anak di Klinik Audiologi RSUD Dr. Soetomo. Derajat keparahan terbanyak adalah profound, dan ditemukan terbanyak bilateral. Faktor risiko yang tidak diketahui terbanyak ditemukan, dan bisa*

merupakan faktor yang berpengaruh pada kejadian lambat bicara dan berbahasa. Temuan ini sesuai dengan rekomendasi program skrining pendengaran yang seharusnya diterapkan pada semua bayi baru lahir.

Kata kunci: *gangguan pendengaran, OAE, BERA, bayi, anak*

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INTRODUCTION

Hearing loss is one of the most common congenital abnormalities, and this disorder is estimated in about 5 infants of every 1000 births. The first three years of life are the most important period for the development of speech and language. As a consequence, infants and children with undetectable hearing loss will miss the golden period of speech and language development, resulting later on delayed and poor proficiency of speech and language, academic achievement, and limited career opportunities.[1,2] This delay in detection will result in the high burden of social costs required to deal with children with hearing loss, as these children need more specialized therapy and special education.[3]

According to the World Health Organization (WHO) in 2007, the prevalence of hearing loss in Indonesia is estimated at 4.2%. The percentage of prevalence of hearing loss in the general population varies from at least 4.2% in Indonesia to 9% in Sri Lanka, 13.3% in Thailand and 16.6% in Nepal. Based on the above figures, there are more than 100 million people suffered from deafness and hearing problems in East Asia. Approximately 50% of children with congenital hearing loss are due to genetic factors. About 50% of the rest are caused by environmental factors such as prenatal or perinatal diseases, or unknown causes.[4] Sense of hearing is required to determine the auditory function of a child with suspected hearing loss. Currently the gold standard for hearing screening examination is otoacoustic emission (OAE) and brainstem evoked response audiometry (BERA).[5]

The OAE examination is an electrophysiological examination to assess objective, automatic (using pass and refer criterion), noninvasive, easy, no time consuming, and practical, so it is very efficient for newborn hearing screening programs, and has a sensitivity of 100% and a specificity of 82 to 87%.[6] While BERA is an examination to assess the hearing function on VIII nerve (auditory nerve) and up to the level of brainstem. The BERA test has a sensitivity of 100%, and a specificity of 97 to 98%. This examination is particularly useful in circumstances where no ordinary hearing test are possible, for example in infants, children with impairments in nature and behavior, low intelligence, multiple defects, and decreased awareness.[5]

The aimed of this study was to obtain a prevalence and description of sensorineural hearing loss in infants and children within two years, 2011 up to 2013 in Dr. Soetomo General Hospital.

METHOD

A retrospective descriptive cross sectional study was performed, with secondary data collected from the medical record in the Audiology Clinic of Dr. Soetomo Hospital. The data included infants and children who had been tested with OAE and BERA within 2 years, in the year 2011-2013. This study revealed the number of patients, age, gender, OAE and BERA examination results, degree of hearing loss, unilateral or bilateral and the risk factors.

RESULTS

The number of all patients that was collected from the medical record of the Audiology Clinic of Dr. Soetomo Hospital in 2011-2013, was 552 patients. From these numbers, it was detected 377 patients (68.3%) with sensorineural hearing loss, consisted of the age range from infants-under-6-months up to 60 months.

The highest sensorineural hearing loss patients were in age group of 12 to 36 months: 237 patients (62.86%), followed by the age group from 36-60 months: 92 patients (24.41%), then from 6-12 months: 32 patients (8.49%), and the lowest age group was under 6 months: 16 patients (4.24%). The patients that were detected with sensorineural hearing loss (SNHL) consisted of 199 males (52.79%), and 178 females (47.21 %) (table 1).

The results of OAE and BERA were shown on table 2. The OAE resulted refer-on 374 patients (67.75%), out of all 552 patients. It was a higher number than the pass result. While on the BERA examination found 366 patients (66.33%). Both OAE and BERA revealed a higher number on refer than pass results.

There were 377 patients (68%), detected with sensorineural hearing loss, out of 552 patients. The degree of hearing loss was shown on table 3, with the highest number on profound group: 329 patients (87.27%). The Auditory neuropathy was found on four patients (1.06%).

The risk factors were identified related with Rubella, CMV infection, history of asphyxia, Hyperbilirubinemia. The highest number remain on unknown risk factor: 310 patients (82.23%). (Tabel. 4)

The sides of hearing loss was shown on table 5, found significantly a higher number on bilateral sensorineural hearing loss: 357 patients (94.69%).

DISCUSSION

The prevalence of hearing loss in Indonesia is estimated at 4.2%.[4] In this study we found 552 patients during 2 years in 2011-2013 and detected 377 patients (68.3%) with sensorineural hearing loss (SNHL). This high prevalence showed as evidence that hearing loss is the most common congenital abnormalities. Since the patients referred to Dr. Soetomo Hospital as the top

Table 1. Characteristic of subject

Characteristic	Category	Number of patients	Percentage (%)
Age (months)	<6	16	4.24
	6-12	32	8.49
	12-36	237	62.86
	36-60	92	24.41
Gender	male	199	52.79
	female	178	47.21
	Total	377	100

Table 2. The result of OAE and BERA examination

Examination	Category	Number of patients	Percentage (%)
OAE	Pass	178	32.25
	Refer	374	67.75
BERA	Pass	186	33.67
	Refer	366	66.33
	Total	377	100

Table 3. The degree of hearing loss with BERA examination

Degree of hearing loss	Number of patient	Percentage (%)
Mild	8	2.12
Moderate	4	1.06
Moderate-severe	9	2.39
Severe	23	6.10
Profound	329	87.27
Auditory neuropathy	4	1.06
Total	377	100

Table 4. Risk factors

Risk factor	Number of patient	Percentage (%)
Rubella infection	14	3.72
CMV infection	15	3.98
Asphyxia history	1	0.26
Hyperbilirubinemia	37	9.81
Unknown	310	82.23
Total	377	100

Table 5. Unilateral or bilateral

Sensory neural hearing loss	Number of patient	Percentage (%)
Unilateral	20	5.31
Bilateral	357	94.69
Total	377	100

referral hospital in Indonesia, it reflected the prominent problem of hearing loss in infant and young children in the community.

The children in age group of 12 to 36 months was found 237 patients (62.86%). (table 2). This is the highest number of age group in infant and young children with SNHL up to 60 months. This finding was relevant with the common reason due to the speech delay, that the parents decided to check the problem and being referred to the audiology clinic. The parents seemed unaware with the appearing of the initial signs of hearing loss until they found a sign of speech delay in older ages of the children. This is a critical period, since the first three years of life is the most important for developing the speech and language.[1]

Early detection and intervention should be accomplished under 6 months of age, but this study revealed it was the lowest percentage: 16 patients (4.24%) (table 1). The Joint Committee of Infant Hearing recommends early detection and intervention for infants with hearing impairment, and implementing the goal of Early Hearing Detection and Intervention (EHDI), so that the delay in speech and communication can be prevented. Its objective is towards the similar development of cognition, reading, and social-emotional with their unimpaired peers.[7]

The Joint Committee on Infant Hearing (JCIH) in the year 2000 issued a standard that says that ages 1 to 3 years is a late age to recognize congenital hearing loss in children.[8] The late diagnose in infants and children will interfere the speech, language, and cognitive development. The most optimal speech development and language habilitation achievement will be obtained from the age

of less than 6 months, while habilitation in aged over 3 years will be less optimal and the prognosis will be poor.[9]

Table 1 showed that the number of SNHL patients in male, 199 (52.79%) and female, 178 (47.21%) with the ratio of 1.12. There was not much difference in the prevalence of hearing loss among male and female, but male has a slightly higher risk of hearing loss.[9]

The degree of hearing loss according to the International Standard Organization (ISO) is divided into normal if the hearing threshold is 0-25 dB, mild >25-40 dB, mild to moderate >40-55 dB, moderate >55-70 dB, severe >70-90 dB, and profound >90 dB.[5] Table 3 showed that most of the SNHL patients in this study was profound as many as 329 patients (87.27%). According to Yussy,[8] a study conducted by Saim as quoted by Asad in Malaysia found that profound degree bilateral SNHL was the highest: 64.7%, followed by severe degree: 16.4%, moderate degree: 16.4%, and mild degree: 2.5%.

Table 4 showed mostly of the risk factors as unknown: 310 patients (82.23%). Another literature that mentioned congenital hearing loss due to genetic factors was about 50%, and the remaining 50% was caused by environmental factors such as prenatal, perinatal or unknown causes. Hearing loss due to genetic factors, was estimated that 70% was non syndromic while 30% was syndromic. In addition, there was mentioned that about 25%-50% of infants with hearing loss did not have risk factors.[10]

A study found that 50% of children with moderate to severe congenital deafness did not have risk factors. This suggests that simply looking at risk factors as an indicator for hearing evaluation would cause 50% of children with congenital deafness to be undiagnosed. Based on these considerations, the current effort to make early detection of hearing loss in infants is determined through the newborn hearing screening (NHS) program.[11]

Table 5 showed a higher number on bilateral SNHL: 357 (94.69%), than unilateral SNHL: 20 (5.31%).

Children with unilateral hearing loss were at risk for further deterioration in hearing, it was about 40% or more of children that first diagnosed with unilateral hearing loss and that about one in six progressed to bilateral hearing loss.[12] The patients with bilateral hearing loss will have difficulty in academic, social, and habitual areas. In the preschool period, patients have developed delayed speech, and it will increase the risk of speech delay development if they are treated after 6 months of age. It is important to identify patients with bilateral or unilateral hearing loss, even if there are no risk factors. The WHO guidelines call for priority for hearing aids and services for children with a mean hearing impairment in the range of 31 to 80 dB HL in the better ear in the frequency range of 500 Hz to 4 kHz, as these are the children who are expected to derive the most benefit from hearing interventions.[12]

The OAE examination in this study resulted refer, in a higher number: 374 patients (67.75%) and also BERA refer: 366 patients (66.3%). These results of OAE and BERA screening were not differ too much in finding the refer. BERA has a higher specificity, there were four patients (1.06%) detected with Auditory Neuropathy (AN). This finding was a special case, even just a small number but it needs a specific approach in its management. AN will result pass with OAE test, because the problem is not in the cochlea, but in the auditory nerve. The electrophysiological examination with automatic results on pass and refer criterion, is practical and has been proven to be very efficient for hearing screening programs to detect the hearing impairment.[5]

Among the SNHL patients with risk factors, found Rubella 14 (3.72%), CMV 15 (3.98%), asphyxia 1 (0.26%), and hyperbilirubinemia 37 (9.81%), but the

highest risk factor was unknown with the number of 310 (82.23%) patients. This result is consistent with other research, that the etiology was unknown for almost half (44.4%): 48 out of 108 of the children.[12] It is important to note that genetic testing should be counted, relating to 50% of children with congenital hearing loss are due to genetic factors.[4]

This study concluded that the majority case was SNHL, that was found in infant and children who came in the Audiology Clinic of Dr. Soetomo Hospital. The degree of SNHL varied with the highest number was profound hearing loss. The unknown of risk factor was the highest number. The sides of hearing loss showed bilateral was much higher than unilateral.

Based on the results above, there was a high burden of SNHL among the patients, with the greater number on profound, bilateral hearing loss, but unknown risk factor. It may relate to the negative impact of the hearing impairment on speech, language, educational, and vocational outcomes in the future.

This study showed a relevance to the recommendation of the Universal New Born Hearing Screening Program (UNHSP) which should be implemented for all new born. Speech and language developmental delay could be prevented with early detection and intervention program, following the UNHSP. Raising awareness and improving access to services at the primary health care level can help to reduce the prevalence and the adverse impact of hearing loss.

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