

Characteristics of Congenital Sensorineural Hearing Loss in Children at ENT Outpatient Clinic Sanglah General Hospital Denpasar in 2017

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Hearing disorders caused by genetic (congenital) and nongenetic (acquired) factors which will affect the patient's communication skills. Joint Committee on Infant Hearing (JCIH) 2000 define hearing disorders in children should be detected at 3 months age and appropriate interventions starting at 6 months age. The intervention by amplification through the installation of hearing aids, cochlear implantation, and special education (speech therapy, special needs school) as a rehabilitation effort, where children are expected to achieve optimal speaking and language skills. This is a retrospective descriptive study by taking secondary data from medical records of patients with congenital sensorineural hearing disorder which underwent BERA examination at Sanglah General Hospital Denpasar in January to December 2017. From 125 children who underwent BERA examination, there were 45 (36%) children with normal hearing and 80 (64%) children with hearing impairments. 22 subjects (31.88%) were in the age group of > 5 years old, with male predisposition. The degree of hearing loss most were very severe as many as 49 children (71.01%) with symmetrical hearing loss in most of the subjects. Most of children with bilateral congenital sensorineural hearing loss (SNHL) are in the age > 5 years old, while men are affected more than women. The degree of hearing loss is mostly very severe, with symmetrical on both ear.

Keywords: Congenital; Hearing Loss; SNHL; Bilateral; Characteristics.

Hearing disorders caused by genetic (congenital) and nongenetic (acquired) factors. Hearing disorders can also arise from birth (prelingual) or after three years old (postlingual) which will affect the patient's communication skills. The incidence of bilateral congenital sensorineural hearing loss is 1-3 / 1,000 births in the normal baby population and 2-4 / 100 babies in intensive care. In West Java with around 37 million population and a population growth rate of 1.8% per year, it is estimated that at least 600 babies are born with severe bilateral sensorineural hearing loss.² Joint

Committee on Infant Hearing (JCIH) 2000 define hearing disorders in children should be detected at 3 months age and appropriate interventions starting at 6 months age. The intervention by amplification through the installation of hearing aids, cochlear implantation, and special education (speech therapy, special needs school) as a rehabilitation effort, where children are expected to achieve optimal speaking and language skills.³ Early detection is very important, especially for children with risk factors for hearing loss, because 50% of newborns have risk factors for hearing problems

since birth. Detection of hearing disorders in children can be done by subjective audiological examination, by paying attention to the child's response to sound in the form of behavioral changes, or by objective examinations, such as brainstem evoked response audiometry (BERA) and auto-acoustic emission (OAE). The BERA examination are objective and reliable it very beneficial for uncooperative patients, infants and children. It can determine the hearing threshold and type of hearing loss objectively. It has a sensitivity of 98% and specificity of 96%. OAE examination is ideal for assessing the integrity of outer hair cells in the cochlea and for screening, because it can be done from one day old babies in a few minutes, but unable to show hearing threshold, only estimating whether the child hears or not, so further examinations needs to be done such as BERA and audiometry.³

METHOD

Research Design

This is a retrospective descriptive study by taking secondary data from medical records of patients with congenital sensorineural hearing disorder which underwent BERA examination at Sanglah General Hospital Denpasar in January to December 2017. The study was conducted at the ENT outpatients clinic Sanglah Hospital Denpasar in January – July 2018.

Sample Research

The study sample was patients with congenital sensorineural hearing disorder at the ENT outpatients clinic Sanglah Hospital Denpasar, from January to December 2017 who meets

Table 1. Characteristics of children with bilateral congenital sensorineural hearing loss (SNHL) based on age

Age (years)	N	%
≤ 1	5	7.25
>1 – 2	12	17.39
>2 - 3	17	24.64
>3 – 4	6	8.70
>4 – 5	7	10.14
>5	22	31.88
Total	69	100

inclusion criteria. The inclusion criteria of this study was all patients diagnosed with bilateral congenital sensorineural hearing disorder. The exclusion criteria were patients with congenital sensorineural hearing disorder with incomplete medical record data and patients with middle and outer ear abnormalities. Sampling was done by consecutive sampling. The sample size is adjusted from the data obtained through medical records during the period January – December 2017.

Data Collection Method

This study uses secondary data from medical records to obtain research data. Data was taken from the medical records of patients with congenital sensorineural hearing disorder who were treated at the ENT outpatients clinic at Sanglah Hospital Denpasar from January to December 2017. The results of the examination were recorded in the data collection sheet for further analysis.

Data Processing

The results of the study were presented descriptively in tables and narratives. Data were grouped according to the variables, then it will be compared according to the variables that have been determined so as to provide a clear and correct illustration.

RESULTS

There were 125 children who underwent BERA examination. There were 45 (36%) children with normal hearing and 80 (64%) children with hearing impairments. Of the 80 children, there were 11 (13.75%) children with hearing impairments that were not included in the study subjects, who has congenital unilateral hearing disorders. The study subjects were 69 (86.25%) children with bilateral congenital hearing disorders.

In table 1 can be seen that of the 69 subjects with bilateral congenital sensorineural hearing loss (SNHL), the most were > 5 years as

Table 2. Characteristics of children with bilateral congenital SNHL according to gender

Gender	N	(%)
Male	40	57.97
Female	29	42.03
Total	69	100

Table 3. Characteristics of children with bilateral congenital SNHL based on degree of hearing impairment

Degree Age (year)	Mild	Moderate	Moderate-Severe	Severe	Very severe	Total
≤1	1	1	1	2	0	5
>1 - 2	0	3	0	2	7	12
>2 - 3	2	1	2	1	11	17
>3 - 4	1	0	0	0	5	6
>4 - 5	0	1	0	0	6	7
>5	0	1	0	1	20	22
Total (%)	4 (5.80%)	7 (10.14%)	3(4.35%)	6 (8.70%)	49 (71.01%)	69 (100%)

Table 4. Characteristics of children with bilateral congenital SNHL based on the character of hearing disorders

Character of hearing impairment	N	(%)
Symmetrical	58	84.06
Asymmetrical	11	15.94
Total	69	100

many as 22 (31.88%) subjects and the least were <1 year as many as 5 (7.25%) subjects.

Table 2 shows predominance of male as many as 40 (57.97%) subjects compared to female as many as 29 (42.03%) subjects.

Table 3 shows the highest hearing impairment degree is very severe, as many as 49 (71.01%) subjects, of which 49 subjects were mostly > 5 years as many as 20 subjects. The least hearing impairment degree were moderate-severe, as many as 3 (4.35%) subjects, which most were > 2-3 years (2 subjects).

Table 4 shows most of the subjects are symmetrical hearing loss, as many as 58 (84.06%) subjects.

DISCUSSION

Bilateral hearing disorders arise from birth (congenital) as many as 69 children. This is consistent with the literature stated hearing disorders in children are often congenital and are generally bilateral sensorineural.

This study obtained most bilateral congenital sensorineural hearing loss (SNHL) in

the age group > 5 years as many as 22 (31.88%) children and least in the age group < 1 year which was only 5 (7.25%) children. When referring to the JCIH standard, the age of diagnosis obtained in this study has not reached the optimal standard because congenital hearing disorders in children must have been detected at < 3 months of age and underwent rehabilitation from the age of 6 months. Delay in diagnosis will disrupt the child's speech, language, and cognitive development. When rehabilitation was done in the age of < 6 month, the most optimal speech and language development will be achieved, in the age of 6 months – 3 years the prognosis is almost the same, but not optimal and in the age of > 3 years has poor prognosis.⁴ This situation may be due to a lack of knowledge of the community, especially parents, of hearing impairment symptoms in children and if not treated early will interfere with the development of speech and language.

In a study by Kalsotra et al., the age of patients diagnosed varied from < 1 month to 22 years (mean age 6.73 years), 40.5% of cases diagnosed before the age of 4 years with a peak age at 4 years (16%).⁵ In a study by Shrivastava and Gupta, the age of patients who came to the ENT clinic are varied between 1 year and 18 years (average age 5, 44 years, SD = 3.43).⁶

The incidence of hearing loss in men was slightly higher than women with a ratio of 1.38 : 1 in this study. Bamiou et al. as cited by Shrivastava⁵ stated male predisposition in children with sensorinural hearing disorders (59.8%). Kalsotra et al. also stated a greater male predisposition of 1.72 : 1 in 261 cases of congenital and acquired hearing disorders consisting of 165 men (63.2%) and 96 women (36.8%).⁵

The study by Shrivastava and Gupta in 50 cases showed 23 men (46%) and 27 women (54%) with a predisposition of 1 : 1,17.6 This correlated with a study by Sanjay et al (2011) which stated almost the same distribution in men and women.7 However, many older studies show more male predispositions. This is likely to occur because increasing awareness of girls than before. However, in this study there was a greater predisposition of men than women, although the comparison was not too large. This is likely due to the fact that in Bali is using patriarchal system which lead to this condition.

This study found mostly severe and very severe hearing loss in all age groups. This is consistent with the literature that the degree of congenital hearing loss is mostly severe and very severe. Study by Saim as quoted by Asad1 in Malaysia showed 2.5% of mild, 16.4% of moderate, 16.4% of severe, and 64.7% of very severe hearing loss.

Symmetrical is more frequent than asymmetrical hearing loss for all degrees of hearing loss. The results of this examination are very important to determine which ears should the hearing aid be placed. In asymmetrical ear hearing loss, the hearing aid are placed in ears with better hearing.

CONCLUSIONS

Most of children with bilateral congenital sensorineural hearing loss (SNHL) are in the age

> 5 years old, while men are affected more than women. The degree of hearing loss is mostly very severe, with symmetrical on both ear.

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