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Hearing loss in pediatric patients with congenital rubella syndrome

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ABSTRACT

Congenital rubella syndrome (CRS) is caused by rubella infection in pregnant women. It was estimated 100.000 children with CRS, with 46% found in developing countries. The CRS consists of symptoms like sensorineural hearing loss (SNHL), congenital heart disease, cataract or congenital glaucoma, and other symptoms. Sensorineural hearing loss is the commonest symptoms compared with others. This study aimed to determine the hearing loss in CRS in Dr. Kariadi Central Hospital, Semarang, Central Java. This was a descriptive study using data from medical records from CRS pediatric patients who had BERA examination from January 2019 until December 2020. The CRS was characterized with one or more symptoms: SNHL, congenital cataract, congenital heart disease, and laboratory IgG and/or IgM Rubella (+). The SNHL was described from refer OAE result, tympanometry A, and BERA with hearing threshold ≥40 dB. Follow-up evaluation was performed with Google form questionnaire. There were 55 CRS cases with 70.9% had bilateral SNHL, mostly with very severe hearing loss. Normal hearing was found in 16 children (28.1%). From 30 children who were followed up, there were 20 children who had bilateral SNHL with 30% were moderate-severe degree and 70% profound. With further multidisciplinary management in children with CRS and parental support, 65% children used hearing aid and 40% had auditory-verbal therapy (AVT). Evaluation of the AVT showed progress in 30% children. In conclusion, SNHL is found in 70.9% children with CRS. Further management using hearing aid and AVT shows progress on few children.

ABSTRAK

Sindrom rubella kongenital (SRK) terjadi akibat adanya infeksi rubella pada ibu hamil. Diperkirakan lebih dari 100.000 anak menderita SRK setiap tahunnya. Sindrom rubella kongenital ditandai adanya *sensorineural hearing loss* (SNHL), cacat jantung bawaan, katarak atau glaukoma kongenital, dan symptom lain. *Sensorineural hearing loss* yang paling umum didapatkan dibanding yang lainnya. Penelitian ini melaporkan kejadian kurang pendengaran pada SRK di RSUP Dr. Kariadi, Semarang. Penelitian deskriptif ini mengambil data rekam medis anak SRK yang diperiksa BERA pada Januari 2019 sampai Desember 2020. Sindrom rubella kongenital ditandai dengan salah satu atau lebih adanya SNHL, katarak kongenital, penyakit jantung bawaan, hasil laborat IgG dan atau IgM rubella (+). SNHL dinilai dari hasil OAE refer, timpanometri A, BERA ambang dengar ≥40dB. Evaluasi tindak lanjut penanganan dengan kuesioner *Google form*. Terdapat 55 (70,9%) anak dengan SRK. Sebagian besar SNHL bilateral berderajat sangat berat. Pendengaran normal dijumpai pada 16 anak (28,1%). Dari 30 anak yang dievaluasi, terdapat 20 anak dengan SNHL bilateral derajat sedang-berat sebanyak 30% dan sangat berat sebanyak 70%. Tindak lanjut dan tatalaksana multidisiplin pada anak SRK disertai dukungan orang tua didapatkan 65% menggunakan alat bantu dengar (ABD) dan menjalani *auditory-verbal therapy* (AVT) rutin (40%). Evaluasi AVT terdapat kemajuan pada 30% anak. Dapat disimpulkan, dijumpai SNHL pada 70,9% anak dengan SRK. Tindak lanjut dan tatalaksana penggunaan ABD dan AVT menunjukkan perbaikan pada sebagian kecil anak.

Keywords:
hearing loss;
congenital rubella syndrome;
habilitation;
auditory-verbal therapy;
hearing aid

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INTRODUCTION

Rubella infection in pregnancy can cause miscarriage, fetal death or congenital abnormalities after birth. Risk level and types of disability depend on gestational age when infected. When the infection of rubella happens in first trimester (12 weeks), about 85% babies had risks to be born with congenital abnormalities; if infection happens in week 13-16 of pregnancy, risks were dropped into 10-20%, while malformation rarely happens after 20 weeks of pregnancy.[1,2] In Indonesia, not many studies on the incidence of rubella are conducted. However, it is estimated that the incidence of rubella infection is quite high. Data from the National Basic Health Research (*Riset Kesehatan Dasar*/Riskesdas) 2011, about 400 cases of congenital rubella syndrome (CRS) were reported, while according to World Health Organization (WHO) in 2012 around 5,000-10,000 babies were born with deafness annually.[3]

Congenital rubella syndrome is characterized with sensorineural hearing loss (SNHL), congenital cataract or glaucoma, congenital heart disease, and developmental delay. Other symptoms are craniofacial anomalies, purpura, and meningoencephalitis. Hearing loss is the most common symptom of CRS. Children with CRS who survive, some will have developmental delay, diabetes mellitus type 1 or thyroiditis.[2,4,5] Some of these impairments can appear or worsen later in the lives of these children. Early introduction and continuation of speech, occupational, physical, and behavior therapies and training with appropriate medical interventions by a multidisciplinary team approach are required to maximize quality of life.[6]

World Health Organization reported more than 100.000 babies were born with CRS worldwide every year and around 46% were reported from southeast Asia including Indonesia. The CRS incidence increased 10 times when epidemic happened.[7] Hearing loss was estimated happened around 80 – 96% in CRS patients and it can be unilateral or bilateral with various degrees in severity.[4] Rubella virus can directly affect cochlea by inducing apoptosis in stria vascularis, cochlear duct and the organ of corti. Stria tissue can be infected and change endolymph structure. Vasculitis occurs and directly damages cells in cochlea, which disrupts myelinization in auditory nerves.[2,8]

Speech and language development are severely affected when hearing impairment is present.[9,10] Auditory–verbal therapy (AVT) is a listening and spoken language (LSL) instructional approach. The AG Bell Academy for Listening and Spoken Language provides certification to specialists who deal with hearing loss children. A previous study revealed that children who participated in AVT can achieve linguistic skills at the same level as their hearing peers.[11] This study aimed to report hearing loss in children with CRS at Dr. Kariadi Central Hospital, Semarang, Indonesia.

MATERIALS AND METHODS

Subjects

This was a descriptive study conducted in Dr. Kariadi Central Hospital, Semarang, Central Java. Data were gathered from medical records from CRS pediatric patients with age 6.2 – 57 months who had BERA examination from January 2019 until December 2020 and met the inclusion and exclusion criteria.

Protocol of study

Initial diagnosis of CRS was made

from medical records if characterized with one or more symptoms i.e. SNHL, congenital cataract, congenital heart disease, and laboratory IgG and/ or IgM Rubella (+). The CRS diagnostic was conducted by pediatricion and examined hearing function was conducted by otolaringologist. The SNHL was scored with refer OAE result, tympanometry A, and BERA with hearing threshold ≥40 dB. Follow-up evaluation was performed by using Google form questionnaire.

Statistical analysis

Data were presented as frequency or percentages and continued by descriptively analysis.

RESULT

Initial data from medical records in period January 2019 until December 2020 showed 55 children with CRS. Boys were found more than girls, mostly aged more than 1 year old. Hearing loss was found in 39 patients (70.9%), all affected bilaterally and 70.9% with very severe hearing loss. The characteristics of subjects are presented in TABLE 1.

Evaluation was performed in September 2021 using Google form, among 30 children with CRS had responses, consisted of 20 CRS children with hearing loss and 10 CRS children with normal hearing. Questionnaire results of 20 CRS children are shown in TABLE 2.

TABLE 1 Basic characteristics of the subjects

Variable	Total [n (%)]
Gender	
▪ Male	32 (59.2)
▪ Female	23 (41.8)
Age of having BERA	
▪ 0 - 1 years old	25 (45.4)
▪ > 1 years old	30 (54.6)
BERA results	
▪ Hearing threshold < 40 dB	16 (29.1)
▪ Hearing threshold ≥ 40 dB	39 (70.9)
▪ SNHL moderate - severe	8 (20.5)
▪ Profound	31 (79.5)
▪ Unilateral	0 (0)
▪ Bilateral	39 (100)
▪ Organ abnormalities	
Hearing loss	14 (35.9)
▪ Hearing loss +1 organ abnormality	19 (48.7)
▪ Hearing loss + > 1 organ abnormalities	6 (15.4)

TABLE 2. Questionnaire results of CRS children (n=30)

Variable	Moderate – severe [n=6 (%)]	Profound [n=14 (%)]	Total [n (%)]
Age (years)			
▪ 0-1	2 (10)	6 (30)	8 (40)
▪ >1	4 (20)	8 (40)	12 (60)
Gender			
▪ Male	3 (15)	8 (40)	11 (55)
▪ Female	3 (15)	6 (30)	9 (45)
Hearing aid type			
▪ Implant	0 (0)	3 (15)	3 (15)
▪ Hearing aid	4 (20)	6 (30)	10 (50)
▪ Without hearing aid	2 (10)	5 (25)	7 (35)
Habilitation			
▪ Speech therapy/AVT	2 (10)	6 (30)	8 (40)
▪ Non-therapy	4 (20)	8 (40)	12 (60)
▪ Therapy result			
▪ No improvement	6(30%)	8(40%)	14(70%)
▪ Improvement	0(0%)	6(30%)	6(30%)

Moderate severe hearing loss was found in 6 children, 4 children used hearing aid, while 2 children only had speech therapy resulted with no improvement. Profound hearing loss were identified in 14 children, 9 children had hearing aid (3 implant, 6 conventional hearing aid device), 6 children had speech therapy resulted with improvement. Parent's occupation and education which supports further management of hearing loss in CRS children (TABLE 3).

TABLE 3. Parent's occupation and education related to hearing loss management

Variable	Implant [n=3 (%)]	Hearing aid [n=10 (%)]	Non Implant/ Hearing aid [n=7 (%)	Total [n=20 (%)
Parent's occupation				
▪ Both parents work	2 (10)	3 (15)	3 (15)	8 (40)
▪ Only father works	1 (5)	7 (35)	4 (20)	12 (60)
Parent's education				
▪ Both parents with scholar degree	3 (15)	4 (20)	1 (5)	8 (40)
▪ Father/mother with scholar degree	0 (0)	3 (15)	3 (15)	6 (30)
▪ Father/mother with non-degree	0 (0)	3 (15)	3 (15)	6 (30)

Parent's occupation and education may affect compliance in follow up control (TABLE 4). Among 20 children with hearing loss (13 children with hearing aid and 7 children with routine speech therapy), 6 children had improve of hearing, 2 children from non hearing aid group, that be conducted on routin speech therapy. All children did no improve of hearing.

TABLE 4. Parent's compliance

Variable	Therapy		Result	
	Routine [n=9 (%)]	Not routine [n=11 (%)]	No improvement [n=14 (%)]	Improvement [n=6 (%)]
Hearing aid type				
▪ Implant	3 (15)	0 (0)	0 (0)	3 (15)
▪ Hearing aid	4 (20)	6 (30)	7 (35)	3 (15)
▪ Without hearing aid	2 (10)	5 (25)	7 (35)	0 (0)
Parent's occupation				
▪ Both parents work	3 (15)	5 (25)	4 (20)	4 (20)
▪ Only father works	6 (30)	6 (30)	10 (50)	2 (10)
▪ Parent's education				
▪ Both parents with scholar degree	6 (30)	2 (10)	5 (25)	3 (15)
▪ Father/ mother with scholar degree	2 (10)	4 (20)	4 (20)	2 (10)
▪ Father/ mother with non-degree	1 (5)	5 (25)	5 (25)	1 (5)

DISCUSSION

Rubella infection in pregnancy can cause miscarriage, stillbirth, congenital abnormalities or asymptomatic infection. It can affect some organs and causes congenital abnormalities which called CRS. Prevalence of CRS in Indonesia is still high. Rubella vaccination can decrease CRS prevalence in developed countries, but can not be conducted completely in some of developing countries including Indonesia.[5] Impact of CRS can cause growth and developmental delay in children, hearing organ impairment can disturb speech and language development, which affect greatly on communication disorder and opportunity on decent education and job.

The result of this study showed that 70.9% children experience bilateral SNHL with mostly profound hearing loss. Age ranged from 6.2 – 57 months. Age 0 – 1 years old were found in 45.4% children and age more than 1 years were found in 56.4% children. A male and female ratio was 1.4:1. It was different with study conducted in Surabaya which found 1.06:1.[12] A study conducted in Bandung reported that 88% of CRS patients experienced hearing loss in which 75% were bilateral. The ratio male and female was 1:1 and mostly occurred in age group of 1-3 months old (22.11%), whereas very severe SNHL were found in children aged 2-14 months old (22%). Furthermore, a study conducted in Tokyo reported that SNHL occurred in CRS patients with ratio of 1:1.[14] Nazme *et al.*[15] reported that SNHL in CRS patients are dominated by male in Bangladesh. A study in Yogyakarta showed severe SNHL in CRS patients were found in 36% children with mostly aged 2-6 months old.[5]

This study also showed hearing loss with and without other organ abnormalities were 70.9%, only hearing loss were 35.9%, hearing loss with one organ abnormality were 48.7%, and hearing loss with more than one organ

abnormalities were 15.4%. This result is similar with the study conducted in Surabaya which reported that bilateral hearing loss was the most common hearing abnormality found with 71.43% cases, hearing loss and congenital heart disease were found in 17.89% cases, hearing loss, congenital heart disease, and ophthalmology abnormalities were found in 16.84% cases, while hearing loss and ophthalmology abnormalities were found in 13.68% cases.[12]

Most CRS study in Indonesia conducted in patients less than 6 months old.[5] The different result of this study compared with other studies may due to still a lot of children who were admitted in Dr. Kariadi Center Hospital, were referred for auditory examination when aged more than 1 years old. Some cases were referred to ENT Departement with complaint of speech delay. The parents realized after their children were enable to talk like the others.

Further follow up and management for CRS patients need multidisciplinary approach involving pediatrician, ophthalmologist, physical medicine and rehabilitation, physicians, and parents support. Parents occupation and education were one of the factors which determines further management of CRS. Parents with good education and socio-economics will get well information about the problem of hearing loss in children and its management. Therefore, they can provided psychosocial and academic development supports of the child and to his or her ultimate quality of life. Children with working fathers and mothers will reduce the focus on children's needs. Among 30 children who gave responses in follow up this study, there were 20 children with hearing loss.

Habilitation with implant or conventional hearing aid device was conducted in 13 children (65%). Eight children had speech therapy/AVT, 6 showed improvement. Success rate of speech therapy for SNHL in children with CRS who used hearing aid only 6 form 13 (46%), 7 children doing to routine speech therapy, 6 children showed hearing improvement, 2 children from non hearing aid group, that be conducted on routin speech therapy. All children no improving of hearing. This may cause by non-routine therapy, parents' commitment and occupation limitation, limitation was cost burden, abnormalities in organs involved and limited time and distance from therapy clinic to home. This result similar with previous study which showed that AVT is an effective intervention option for the AVT group.[16]

CONCLUSION

Hearing loss in CRS patients are commonly found (70.9%). Habilitation with speech therapy/AVT conducted in some children shows improvement in listening and speech. These findings support the positive effect of creating an appropriate educational environment by considering individualized needs. Also, exploring parental needs is very important for planning and making decisions in the rehabilitation process.

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