CHARACTERISTICS OF HEARING IMPAIRMENT IN PATIENTS WITH SUSPECTED CONGENITAL RUBELLA SYNDROME AT THE ENT CLINIC OF PROF. dr. I.G.N.G. NGOERAH HOSPITAL DENPASAR, INDONESIA

I Gede Wahyu Adi Raditya[1], I Made Wiranadha

Department of Otorhinolaryngologists Head and Neck, Faculty of Medicine, Udayana University, Denpasar, Indonesia

ABSTRACT

Asymptomatic rubella virus infection can cause delays in the diagnosis of rubella in pregnant women, leading to the development of congenital rubella syndrome (CRS). The clinical manifestations of congenital rubella syndrome are called the rubella triad, which includes heart problems, eye problems, and hearing disorders. Sensorineural hearing loss is the most common type of hearing loss in children with rubella infection. This study aimed to determine the characteristics of hearing loss in CRS patients. This study used a retrospective descriptive research design, utilizing secondary data from the medical records of patients with CRS who underwent hearing examinations at the Ear, Nose, Throat (ENT) Polyclinic of Prof. dr. I.G.N.G. Ngoerah Hospital Denpasar, Indonesia. The study found that the most common age group among suspected CRS patients was 6 - 12 months (51.42%), the majority of these patients were male (62.86%), the most frequent clinical signs and symptoms in suspected CRS patients were congenital heart disease (CHD) as a major criterion (60%), and microcephaly as a minor criterion (77.14%). Based on the CRS case criteria, none of the suspected CRS patients were diagnosed with confirmed CRS, with most cases being classified as clinical CRS (65.71%). The hearing loss in suspected CRS patients was entirely sensorineural type with the majority of cases being bilaterally and to a very severe degree (53.57%)

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Corresponding author
I Gede Wahyu Adi Raditya
✉ dr.gedewahyu@unud.ac,id
Department of ORL-HNS, Faculty of Medicine, Udayana University, Denpasar, Bali

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INTRODUCTION

Rubella is a contagious RNA virus infection and there is no specific treatment for this infection. In 2016, there were 22,361 cases reported worldwide[1]. A survey conducted in Indonesia from 2010 to 2015 estimated that there were 30,436 cases of rubella. Surveillance data over the last five years shows that 70% of rubella cases occurred in the <15-year age group. Additionally, a study on the estimated burden of congenital rubella syndrome (CRS) in Indonesia in 2013 estimated 2,767 cases of CRS, with an incidence of 82 per 100,000 among mothers aged 15-19 years and decreasing to 47 per 100,000 among mothers aged 40-44 years[1]. Rubella

infection during pregnancy can lead to miscarriage, stillbirth, and CRS in neonates, especially during the first trimester of pregnancy[1].

Asymptomatic rubella virus infection can delay the detection of the virus in pregnant women, which may lead to CRS. The clinical manifestations of CRS known as the rubella triad, include heart defects, eye problems, and hearing disorders[2]. Sensorineural deafness is the most common type of hearing loss in children with rubella infection[3]. Parents often realize that their child has congenital deafness when the child is older, typically between 2-5 years of age, as growth and development, particularly speech and language abilities, may seem normal in the early years[4]. Children with congenital deafness may experience speech delays, language disorders, social behavior deviations, and decreased cognitive abilities[5].

The incidence of congenital bilateral sensorineural hearing loss in Indonesia is 1–3 per 1,000 births in the normal baby care population and 2–4 per 100 babies in intensive care. In West Java, with a population of approximately 37 million and a population growth rate of 1.8% per year, it is estimated that at least 600 babies will be born with severe bilateral sensorineural hearing loss each year[6].

In developing countries, early detection of hearing loss is not yet fully optimized[7-9]. Early detection is crucial, particularly in children with risk factors for hearing loss, as 50% of newborns with these risk factors experience hearing loss from birth. Hearing disorders in children can be detected through subjective audiological examination, which involves observing the child's behavioral response to sounds, or through objective examinations such as Brainstem Evoked Response Audiometry (BERA) and Otoacoustic Emission (OAE)[4]. Given the high number of CRS cases and the public's limited awareness of hearing loss associated with CRS, the authors conducted a study on the characteristics of hearing loss in patients with suspected congenital rubella syndrome at the ENT clinic at Prof. dr. I.G.N.G. Ngoerah Hospital in Denpasar.

MATERIALS AND METHODS

This study used a retrospective descriptive research design, utilizing secondary data from medical records of patients with suspected CRS who underwent hearing examinations at the ENT clinic of Prof. dr. I.G.N.G. Ngoerah Hospital, Denpasar, during the period January to December 2018. The research samples consisted of patients with suspected CRS who underwent hearing examinations at the ENT clinic, Prof. dr. I.G.N.G. Ngoerah Hospital, Denpasar in January – December 2018. Ethical clearance for this research was obtained from the Research Ethics Commission No. 1119/UN14.2.2.VII.14/LT/2020 of the Prof. dr. I.G.N.G. Ngoerah Hospital.

Sampling was conducted using the total sampling method, where all data from medical records that met the study's criteria were included in the analysis. The inclusion criteria for this study were all patients with suspected CRS who underwent hearing examination, while the exclusion criteria included patients with suspected CRS who had incomplete medical record data, patients with external and middle ear abnormalities, and patients who had not completed all CRS surveillance examinations.

RESULT

Sample selection was conducted using the total sampling method, resulting in 35 samples. Based on age, the majority of patients with suspected CRS were examined at 6 - 12 months of age, accounting for 18 patients (51.42%), followed by 12 - 18 months with 11 patients (31.42%) and 18 - 24 months with 6 patients (17.16%). Among the 35 research samples, 22 patients (62.86%) were male and 13 patients (37.14%) were female. The sample characteristics are presented in Table 1.

Table 1. General characteristics of the study sample based on age at the time of hearing examination

Variables	Total	Percentage (%)
Age		
<6	0	0
6 – 12	18	51.42
>12 – 18	11	31.42
>18 – 24	6	17.16
>24	0	0
Sex		
Male	22	62.86
Female	13	37.14

Based on the clinical signs and symptoms of CRS, the samples were grouped according to the presence or absence of symptoms and signs. Major criteria included cataracts, congenital glaucoma, congenital heart disease, and retinal pigmentation. Minor criteria included purpura, splenomegaly, jaundice, microcephaly, mental retardation, meningoencephalitis, and radiolucent bone disease. As shown in Table 2, congenital heart disease (CHD) was the most common clinical sign and symptom according to the major criteria, found in 21 samples (60%). In terms of minor criteria, microcephaly was the most frequently observed present in 27 (77.14%) samples.

Based on the criteria for CRS cases, 23 (65.71%) samples were classified as clinical CRS, 3 (8.57%) samples were classified as congenital rubella infection, 9 (25.72%) samples were not classified as CRS, and none of the samples were diagnosed as confirmed CRS cases.

Based on the results of the OAE examination, the highest number of referral results were obtained for both the right and left ears. Specifically, 20 (57.14%) samples received referral results for the right ear, and 21 (60%) received referral results for the left ear (Table 4).

Table 5 presents the tabulated results of the characteristics of the research sample based on the location of hearing loss, categorized as bilateral or unilateral. Of the 35 samples, 7 (20%) had no hearing loss in either ear, 4 (11.43%) had hearing loss in one ear, and 24 (68.57%) had hearing loss in both ears.

Table 6 shows the characteristics of the research sample based on the degree of hearing loss experienced. Among the 4 patients with unilateral hearing loss, the majority had hearing loss in the left ear, with the highest degree being very severe in 2 (50%) samples. Among the 24 patients with bilateral hearing loss, the most severe degrees were observed in both ears. Specifically, 15 (62.5%) samples had very severe hearing loss in the right ear, and 13 (54.17) samples had very severe hearing loss in the left ear. Overall, the most common degree of hearing loss experienced by patients with suspected CRS was very severe, found in 30 (53.57%) samples.

Table 2. Characteristics of the sample based on clinical signs and symptoms

Clinical signs and symptoms	Yes	No	Total
Major Criteria			
Congenital Cataract	11 (31.42%)	24 (68.57%)	35 (100%)
Congenital Glaucoma	0 (0%)	35 (100%)	35 (100%)
Congenital heart	21 (60%)	14 (40%)	35 (100%)
disease Retinal pigmentation	0 (0%)	35 (100%)	35 (100%)
Minor criteria			
Microcephaly	27 (77.14%)	8 (22.86%)	35 (100%)
Jaundice <24 jam	2 (5.71%)	33 (94.29%)	35 (100%)
Purpura	4 (11.43%)	31 (88.57%)	35 (100%)
Splenomegaly	1 (28.56%)	34 (97.14%)	35 (100%)
Mental retardation	2 (5.71%)	33 (94.29%)	35 (100%)
Meningoencephalitis	0 (0%)	35 (100%)	35 (100%)
Radioluscent Bone Disease	0 (0%)	35 (100%)	35 (100%)

Table 4. Characteristics of the research sample based on the results of the OAE examination

	Right		Left	
	N	(%)	N	(%)
pass	15	42.86	14	40
refer	20	57.14	21	60
	35	100	35	100

Table 5 Characteristics of the research sample based on the location of the hearing loss

Location of Hearing Loss	Total	Percentage (%)
Normal	7	20
Unilateral	4	11.43
Bilateral	24	68.57
Total	35	100

Table 6 Characteristics of the research sample based on the degree of hearing loss

	Unilateral		Bilateral		Total ear
	Right	Left	Right	Left	
Normal	3 (75%)	1 (25%)	0 (0%)	0 (0%)	4 (7.14%)
Light	0 (0%)	1 (25%)	2 (8.33%)	0 (0%)	3 (5.36%)
Moderate	0 (0%)	0 (0%)	4 (16.67%)	8 (33.33%)	12 (21.43%)
Moderately Severe	1 (25%)	0 (0%)	1 (4.17%)	2 (8.33%)	4 (7.14%)
Severe	0 (0%)	0 (0%)	2 (8.33%)	1 (4.17%)	3 (5.36%)
Profound	0 (0%)	2 (50%)	15 (62.5%)	13 (54.17%)	30 (53.57%)
Total	4 (100%)	4 (100%)	24 (100%)	24 (100%)	56 (100%)

DISCUSSION

In this study, it was found that, based on age, the patients with the highest suspicion of CRS were examined at 6 - 12 months of age, with 18 patients (51.42%), followed by 12 - 18 months of age with 11 patients (31.42%), and 18 – 24 months of age with 6 patients (17.16%). This finding was in contrast with research conducted by Apriliani in 2017 at Wahidin Sudirohusodo Hospital, Makassar, which reported that 15 (50%) samples were examined at ages older than 24 months (2 years)[9]. Additionally, Dammeyer's 2010 study in Denmark found that 100% of patients with suspected CRS had their hearing checked before the age of 1 year[3]. The earlier age of examination in this study may be attributed to the effective CRS surveillance program implemented by the Indonesian Ministry of Health at Prof. dr. I.G.N.G. Ngoerah Hospital Denpasar, which allows for earlier detection and intervention for patients with suspected CRS.

Of the 35 research samples, CRS occurred more frequently in male patients,

with 22 patients (62.86%) compared to 13 female patients (37.14%). According to the CRS surveillance report by the Indonesian Ministry of Health in 2017, no significant differences between males and females[2]. Similarly, studies by Aprilia in 2017 and Dammeyer in 2010 also reported no significant differences between males and females[3,9].

A range of clinical signs and symptoms characterizes congenital rubella syndrome[10]. Major signs and symptoms include cataracts or congenital glaucoma, congenital heart disease, retinal pigmentation, and sensorineural hearing loss. Minor signs and symptoms include purpura, splenomegaly, jaundice appearing within 24 hours of birth, microcephaly, mental retardation, meningoencephalitis and radiolucent bone disease[11]. In this study, congenital heart disease (CHD) was the most common major clinical sign, found in 21 samples (60%). Among the minor criteria, microcephaly was the most frequent finding, observed in 27 (77.14%) samples. This finding aligns with Dammeyer's 2010 research, which also identified CHD as the most common clinical finding, followed by sensory neural deafness[3]. Meanwhile, Apriliani's 2017 research found microcephaly to be the most common minor sign, present in 18 out of 30 research samples[9]. Additionally, Ezike, et al. (2013) found that CRS signs are related to the gestational age at which the mother was infected with rubella, reflecting the stage of fetal development during the infection[6,12]

Based on the criteria for CRS cases, this study found that 23 (65.71%) samples were classified as clinical CRS, 3 (8.57%) samples had congenital rubella infection, 9 (25.72%) samples were not classified as CRS, and none of the samples were diagnosed as confirmed CRS case. The number of suspected CRS cases identified in 2018 was higher (35 cases) compared to 2017 (20 cases) and 2016 (22 cases). In 2017, there was 1 confirmed CRS case with 70% of cases being clinical CRS, while in 2016, 64% of cases were clinical CRS. In Indonesia, not all hospitals conduct CRS surveillance. For instance, Apriliani's 2017 study did not identify any confirmed CRS cases among 30 patients with suspected CRS at Wahidin Sudirohusodo Hospital in Makassar[9].

Based on the results of the OAE examination, the highest number of referral results were observed for both ears. Specifically, 20 samples (57.14%) received referral results for the right ear, and 21 samples (60%) received referral results for the left ear. The OAE examination is a standard screening tool for detecting hearing loss in CRS surveillance[13]. If a baby under 1-year-old who is at high risk shows OAE referral results in one or both ears, the patient is immediately categorized as a suspect for CRS[14]. In contrast, for patients not considered high risk, a follow-up examination is conducted 1 month later[15].

Of the 35 samples in this study, 7 (20%) had no hearing loss in either ear, 4 (11.43%) had hearing loss in one ear, and 24 (68.57%) had hearing loss in both ears. All samples with hearing loss had sensorineural hearing loss. Ezike et al. (2013), reported similar findings, where all patients with suspected CRS had sensorineural deafness, with 87.6% of cases being bilateral[6,8,16]. This is consistent with the pathogenesis of rubella virus-related hearing loss, where the viral infection causes cochlear damage and direct cell death in the organ of Corti and stria vascularis, typically affecting both

ears[7,17,18].

Among the 4 patients with unilateral hearing loss, the majority had hearing loss in the left ear, with the highest degree being very severe in 2 samples (50%). Of the 24 samples with bilateral hearing loss, the most severe degrees were observed in both ears. Specifically, 15 samples (62.5%) had very severe hearing loss in the right ear, while 13 samples (54.17%) had very severe hearing loss in the left ear. Overall, the most common degree of hearing loss in patients with suspected CRS was very severe, found in 30 samples (53.57%). This finding aligns with research by Kandari et al. (2016), which found that 63% of children undergoing BERA examination were profoundly deaf[8]. There are relatively few studies examining the degree of hearing loss in infants, particularly those with suspected CRS, due to the limited use of ASSR and BERA as diagnostic modalities[8,19,20].

CONCLUSION

This study found that the most common age for patients with suspected CRS was 6-12 months, and the majority of these patients were male. The most common clinical signs and symptoms were congenital heart disease (CHD) according to the major criteria and microcephaly according to the minor criteria. Based on CRS case criteria, none of the patients with suspected CRS were diagnosed with confirmed CRS; most were classified as clinical CRS. All patients with suspected CRS exhibited sensorineural hearing loss, with the majority experiencing bilateral and very severe (profound) hearing loss.

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CONFLICT OF INTEREST

There are no conflicts of interest

ETHICS CONSIDERATION

The research received ethical clearance from the Prof. dr. I.G.N.G. Ngoerah Hospital. The approval date is May 28th, 2020, with number 1119/UN14.2.2.VII.14/LT/2020

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AUTHOR CONTRIBUTION

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