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# Cochlear implants evaluation for congenital rubella syndrome patients in Dr. Soetomo Academic Medical Center

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***Abstract*---**Congenital rubella syndrome (CRS) is an infection caused by rubella virus that transmitted to the fetus during pregnancy which can cause congenital hearing loss. Cochlear implant is an effective therapy in children with severe to profound bilateral hearing loss. Categories of Auditory Performance (CAP) method is a receptive scale assessment of auditory stimulation that can be applied to all children regardless of age, before and after cochlear implant. Evaluating the results of cochlear implants using CAP-II in patients with CRS. A retrospective descriptive study were conducted to samples taken from medical records of CRS patients who had cochlear implants in 2011-2018 at ORL-HNS department of Dr. Soetomo Academic Medical Center. Evaluation of CAP-II results was performed 6 months, 12 months and 24 months after cochlear implant. Eleven CRS patient who underwent cochlear implant during 2011-2018. Six months after cochlear implant showed the highest scale was 6. At 12-months evaluation showed the highest scale was 7, while at 24-months evaluation showed the highest scale was 8. Cochlear implants provide good auditory performance based on CAP-II scale in sensorineural hearing loss of CRS patients.

***Keywords*---**congenital rubella syndrome, cochlear implant, CAP-II, infectious disease.

## Introduction

Congenital hearing loss is a hearing disorder occurs at birth that can be caused by genetic or environmental factors. One of the important causes of congenital hearing loss is infection caused by rubella virus. Rubella (German Measles) is a viral infection caused by rubella virus (RuV) to the fetus during pregnancy. Rubella infection in pregnant women can cause fetal death or congenital disorders known as congenital rubella syndrome (CRS) in newborns. This infection causes various abnormalities such as hearing loss, balance disorders, congenital cataracts, heart defects, motor disorders, cerebral lesions with late onset complications such as thyroid disorders, diabetes, growth hormone deficiency and panencephalitis (Korver, et al., 2016; Sao&Navya, 2017).

World Health Organization (WHO) estimates that there are 100,000 children born with CRS every year. Studies showed the greatest frequency of congenital infections (80%) occurs during the first trimester, and a smaller risk at the end of the second trimester. Sensorineural hearing loss is the most common complication found in children with CRS. Histopathological studies reported inner ear is the most common disorder during 6-11 weeks of pregnancy. Severe hearing loss is more common compared than mild or moderate hearing loss (Sao&Navya, 2017). Hearing loss cause disorder at speech, language and cognitive development. Early detec tion of CRS cases is needed to collect for determining strategy of CRS prevention (Rosalina&Purnami, 2020; Wahyuni&Ekorini, 2013).

General management of hearing loss includes hearing aids, cochlear implants and bone anchored hearing aid (Korver, et al., 2016). Cochlear implant is an electronic device implanted to the cochlea which converts mechanical energy into electrical energy then delivered through the auditory nerves (Ekorini, 2013). Cochlear implants is reported as an effective therapy for children with severe to profound bilateral hearing loss and is safe for children aged 12 months (Chen&Oghalai, 2016). There are various methods to evaluate the development of a child after cochlear implant. One of the methods which is internationally recognized to assess the development of auditory perceptions using the Categories of Auditory Performance (CAP) method. The CAP method is a receptive scale assessment of auditory stimulation that can be applied to all children regardless of age, before and after implantation. The CAP method is currently being developed into CAP-II with more assessment scores (Arief, et al., 2018). This study aims to evaluate cochlear implant results in patients with congenital rubella syndrome in Dr. Soetomo Academic Medical Center from 2011 to 2018.

## Methods

A retrospective descriptive study was conducted in 11 samples of CRS patients from 131 patients with hearing loss who underwent cochlear implants during 2011 to 2018 in the Neurotology Division of the Otorhinolaryngology-Head and Neck Surgery (ORL-HNS) in Dr. Soetomo Academic Medical Center. The evaluation of CAP-II results after cochlear implant was assessed by Neurotologist consultant of ORL-HNS specialist. The diagnosis of CRS was based on a positive result of anti-rubella Immunoglobulin-G (IgG) and/or Immunoglobulin-M (IgM) test. The sample data were divided based on sex, age at identification, age at

implant, anti-rubella serology test, magnetic resonance imaging (MRI) and computed tomography (CT) scan, other congenital abnormalities and CAP-II evaluation results at 6 months, 12 months and 24 months after cochlear implant.

**Results**

The prevalence of rubella sample was 11 (8,4%) of 131 hearing loss patients who underwent cochlear implant. The sample were evaluated after cochlear implant using Category of Auditory Performance-II (CAP-II) at 6 months, 12 months and 24 months later.

**Demographic Data**

Demographic data in this study were divided based on sex and age. The age distribution was divided into age at identification and age at implant.

Table 1. Demographic data

| | Amount | Percentage |
|---|---|---|
| Sex | | |
| Female | 6 | 54,5% |
| Male | 5 | 45,5% |
| Age at identification (month) | | |
| <12 | 10 | 90,9% |
| 12 – 24 | 1 | 9,1% |
| Age at implant (month) | | |
| 12 – 24 | 5 | 45,4% |
| 25 – 36 | 2 | 18,2% |
| 37 – 48 | 2 | 18,2% |
| >48 | 2 | 18,2% |
| Total | 11 | 100% |

The data of CRS patients sex in this study with a ratio 1.2:1 and mean age at identification was 6.2 month. The youngest age at cochlear implant was 13 month and the oldest was 54 month with mean age at cochlear implants was 24.6 month.

**Clinical Data**

The clinical data in this study were divided based on anti-rubella IgM and IgG serology test, MRI and CT scan. Factors that determine the success of cochlear implants consist of heart defect, eye disorder and motor disorder.

Table 2. Anti-rubella serology tests

| Result | Amount | Percentage |
|---|---|---|
| IgM(+) IgG(-) | 1 | 9,1% |
| IgG(+) IgM(-) | 2 | 18,2% |
| IgM(+) IgG(+) | 8 | 72,7% |
| Total | 11 | 100% |

Serology test in 11 patients showed the highest number was 8 patients with positive IgM and IgG, followed by 2 patients with positive IgG and 1 patient with positive IgM.

Table 3. MRI and CT scan results

| Results | Total | Percentage |
|---|---|---|
| Normal | 2 | 18,2% |
| Periventricular Leukomalacia | 7 | 63,6% |
| Other disorders | 2 | 18,2% |
| Total | 11 | 100% |

The results of MRI and CT scan evaluation in CRS patients obtained 7 patients with periventricular leukomalacia, normal result was found in 2 patients, and other disorders in 2 patients consisting of one patient with multiple small lesions in the frontal lobe and one patient with mild mastoiditis on the right side.

Table 4. Other congenital abnormalities

| Characteristics | Total | Percentage |
|---|---|---|
| Hearing loss only | 1 | 9,1% |
| Heart defect | 5 | 45,4% |
| Eye disorder | 6 | 54,5% |
| Motor disorder | 8 | 72,7% |

Table 4 showed that only 1 of the 11 samples experienced hearing loss. Heart disorder was found in 5 patients, eye disorder in 6 patients, and motor disorder was found in 8 patients.

**Evaluation After Cochlear Implant**

Evaluation after cochlear implant in this study used the CAP-II scale, at 6 months, 12 months and 24 months after cochlear implant.

Table 5. CAP-II after cochlear implant

| Patient | 6 months | 12 months | 24 months |
|---|---|---|---|
| Patient 1 | 1 | 2 | 4 |
| Patient 2 | 4 | 4 | 5 |
| Patient 3 | 1 | 3 | 4 |
| Patient 4 | 1 | 2 | 2 |
| Patient 5 | 6 | 7 | 8 |
| Patient 6 | 5 | 6 | 8 |
| Patient 7 | 5 | 7 | 8 |
| Patient 8 | 1 | 1 | 2 |
| Patient 9 | 5 | 6 | 7 |
| Patient 10 | 1 | 1 | 1 |
| Patient 11 | 3 | 3 | 4 |

Table 5 showed the highest CAP-II value at 6 months after the cochlear implant was scale 6 (1 patient), and the lowest number was scale 1 (5 patients). At 12 months evaluation, the highest number was on category 7 (2 patients). At 24 months evaluation, the highest number was at scale 8 as many as 3 patients, and there were still patients with scale 1 as many as 1 patients.

**Discussion**

Rubella virus infection occurs in early pregnancy cause a serious multi-organ disorder in babies called congenital rubella syndrome (CRS). Babies born with rubella can develop malformations such as hearing loss, congenital cataract, congenital heart defects, acute meningoencephalitis and panencephalitis. The risk of intellectual disability and behavioral disorders is also increased in children with CRS (Wondimeneh, et al., 2018). Sensorineural hearing loss in newborns with CRS can be caused by damage to sensory organs of hearing such as the organ of Corti (Sao&Navya, 2017). The ratio of females with CRS to males in this study was 1.2:1. This finding was consistent with a study conducted by Ciscare et al in 2017 with similar results, 51,6% for females and 48,4% for males (Ciscare, et al., 2017). The different results were obtained by other research, male mostly found than female with ratio 1,2:1 (Paramita&Purnami, 2020)

This study showed that the most of CRS patients were detected at the age of less than 12 month with an average age at identification of 6.2 month. This age classification is based on the ability to speak in normal children without hearing loss. The results showed delays in diagnosing hearing loss and hearing screening in newborns had not been implemented. Hearing loss according to The Joint Committee on Infant Hearing (JCIH) is diagnosed and intervened before the age of 6 month. Early detection of all newborns should be done at 1 month of age, diagnosed at 3 month and intervened at 6 month of age. Babies born with sensorineural hearing loss and hearing loss before the age of 4 will forget all memories and speech. Children detected and intervened before 6 month of age differed significantly in language proficiency compared to children detected and intervened after 6 month of age (Ekorini, 2016).

The mean age at cochlear implant was 24.6 month. Several studies suggested that the age at implant affects the hearing and language. The earlier implant will result in better development. Many children implanted before 2 years old have similar speech and language skills as children their age who do not have hearing loss (Zhou, et al., 2013). This study showed the most patients had positive anti-rubella IgM and IgG. Study conducted by Yudistira et al reported 95.7% hearing loss on positive IgM serology tests. Board et al quoted by Yudistira reported that hearing loss occurred in 48.5% of all samples with positive serology results. Some of the infants had positive IgM result at the of 0-3 months and then at the age 4-6 months the infants had positive IgG results (Kesuma, et al., 2020)

The patients in this study who had pathological findings on MRI and CT scan were still given cochlear implants because they did not contraindicate cochlear implants. Contraindications to cochlear implants include absence of the cochlea or cochlear nerve, and severe mental impairment or mental retardation. Relative contraindications are acute or chronic otitis media and mastoiditis without

eradication, general weak condition, uncontrolled epilepsy, and unable to carry out rehabilitation after surgery (Egilmez&Kalcioglu, 2015). Periventricular leukomalacia (PVL) is one of the brain injuries in infants in the form of focal necrosis and gliosis in the periventricular white matter. The study conducted by Shang et al reported that 82 (45.3%) of 181 PVL cases had TORCH infection in the prenatal period. The manifestations of PVL are often associated with epilepsy, visual and hearing problems, mental retardation and impaired language development. Periventricular leukomalacia can affect the degree of disability and quality of life in children. Literature stated that the prevalence of vision and hearing problems in PVL is 26.96% (Shang, et al., 2015).

This study found other symptom than sensorineural hearing loss like congenital hearing loss and eye disorder. Journal reported by Absalem et al stated that sensorineural hearing loss occured in 58% of CRS patients. The study stated that about 40% of CRS patients can experience only hearing loss as the main disorder without other manifestations. Eye disorder occurred in 43% of CRS patients and 80% had bilateral eye disorders. Congenital heart defects such as patent ductus arteriosus (PDA) and pulmonary artery stenosis occured in 50% of infected children by the second month of pregnancy. Hearing loss and heart defects occured in all infected infants in the first 10 weeks of pregnancy, while hearing loss could occur in weeks 13-16 of pregnancy (Absale, et al., 2017).

There are various methods to evaluate the development of children after cochlear implants. One method of monitoring children after cochlear implants that is internationally recognized and easy to apply is to assess auditory deevelopment using Categories of Auditory Performance (CAP) method (Arief, et al., 2018). CAP assessment is a scale of auditory perception ability ranging from 0 to 7. Two categories (categories 8 and 9) have been added to the CAP scale to increase the result value of implants, hereinafter referred to as CAP-II. The Categories of Auditory Performance assesses children's responses to everyday situations, at home and school (Gilmour, 2010; Absalem, et al., 2017).

This study found 1 patient with CAP-II category 6 at 6 months after cochlear implant. The 12-month evaluation after implant obtained 2 patients with CAP-II category 7. The 24-month evaluation found 3 patients with category 8. Patients who still had scale 1 at 24 months after cochlear implants were caused by other disorders such as congenital cataracts, motor and cognitive disorders. Lyu et al reported that CAP values in children with hearing loss under 3 years after cochlear implants showed very rapid and significant changes at 6 months after implant, therefore listening practice in the first 6 months was crucial. At 6 to 24 months after implant showed an increase in the ability to hear significantly and quickly, although it was slower than at the first 6 months, and entered a plateau phase at 24 months. Children with cochlear age between 2-4 years taken 3 years to reach a normal CAP value, whereas it is very difficult for children with cochlear aged >4 years to reach normal CAP values (Lyu, et al., 2019).

Research conducted by Zhou et al also used CAP to assess the speech perception ability of children with cochlear implants. This evaluation measured supraliminal abilities which reflect everyday listening skills realistically. Post implantation CAP values showed an improvement during the follow-up period.[12] A case report

conducted by Pramila et al in 2018 in India reported a child with CRS who had severe hearing loss accompanied by heart defects, retinopathy, motor and cognitive disorders underwent cochlear implants with good results. Patients have a cochlear implant at age 5 with the results of being able to detect sound, sound perception and be able to communicate well (Pramila, et al., 2018).

## Conclusion

Cochlear implants provide good auditory performance based on CAP-II scale in sensorineural hearing loss of CRS patients. Cochlear implants can be given as early as possible after being diagnosed with sensorineural hearing loss. Further research is needed to compare the results of CAP-II before and after cochlear implants in CRS patients.

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