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(RESEARCH ARTICLE)

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Hearing loss in children with congenital heart disease

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Abstract

Background : Congenital heart disease (CHD) is a dangerous disease, about 50% of deaths will occur in the first month of life. In developed countries almost all types of congenital heart disease have been detected in infancy even at less than 1 month of age, whereas in developing countries many are only detected after the child is older, so in some severe CHD types may have died before being detected. Rubella infection in pregnancy or congenital rubella syndrome (CRS) can cause miscarriage, fetal death, or congenital abnormalities after birth. CRS is characterized by sensorineural hearing loss (SNHL), congenital cataracts or glaucoma, congenital heart disease, and developmental delays.

Purpose : This case report was submitted in order to find out how to properly treat patients with hearing loss with sensory deafness and congenital heart disease.

Case : Reported a 3-year-old 6-month-old boy diagnosed with very severe degree sensorineural deafness and congenital heart disease based on anamnesis, physical examination and examination of OAE, BERA and ASSR for examination of auditory function) and echocardiography examination for examination of cardiac function.

Conclusion : From the history, physical examination and examination of OAE, BERA, ASSR showed patients with very severe sensorineural hearing loss of the left and right ear and congenital heart disease, planned use of hearing aids or cochlear implantation and cardiac consul for CHD treatment.

Keywords: CHD; Sensorineural hearing loss; OAE; BERA and ASSR

1. Introduction

Congenital deafness is hearing loss that is present from birth, both genetic and non-genetic and is usually sensorineural in nature. This condition occurs as a result of prenatal, natal and postnatal risk factors.[1]

Rubella infection in pregnancy or Congenital rubella Syndrome (CRS) can cause miscarriage, fetal death, or congenital abnormalities after birth. CRS is characterized by sensorineural hearing loss (SNHL), congenital cataracts or glaucoma, congenital heart disease, and developmental delays. Other symptoms are craniofacial abnormalities, purpura, and meningoencephalitis. Hearing loss is the most common symptom of CRS.[2,3]

Hearing loss in children needs to be detected as early as possible considering the important role of hearing function in the process of speech development. Identify hearing loss early by observing the child's reaction to sounds or testing hearing function using simple methods and equipment. Currently, there are many methods for assessing children's hearing function. Hearing tests on children cannot be postponed just because the child's age does not allow for a hearing test. Without a hearing screening program, hearing loss is only discovered at the age of 18 – 24 months.[4]

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Delays in a child's language skills can be identified if we know whether the child's current abilities are appropriate for his current age or not. Knowledge of language skills in children is the key to early detection if there is a delay. We can assess based on milestones in a child's language development.

The 2019 Joint Committee on Infant Hearing (JCIH) emphasized early hearing detection and intervention (EHDI) through screening for all newborns aged 1 month. Early detection of hearing loss is very important to avoid delays in diagnosis and intervention so that it can improve children's speech and language development in the future.[5]

Early diagnosis and intervention are important to acquire hearing, speech, and linguistic skills thereby contributing to positive development in children. Newborn hearing screening programs have been introduced to facilitate early identification of hearing loss in children and allow timely intervention through counselling, hearing aids, or cochlear implants in severe cases.[6]

Congenital heart disease (CHD) is a dangerous disease, around 50% of deaths will occur in the first month of life. In developed countries almost all types of congenital heart disease are detected in infancy even at less than 1 month of age, whereas in developing countries many are only detected after the child is older, so that some types of severe CHD may have died before being detected.[7]

Indonesia's population is 200 million, and the birth rate is 2%, then the number of CHD sufferers in Indonesia will increase by 32,000 babies every year. The main obstacle in treating children with CHD is the high cost of examinations and operations. Our experience at the RSCM Cardiology Polyclinic is that the majority of children with CHD who seek treatment come from poor families.[8]

Hearing function influences speech and language development, socialization and cognitive development. So it is important to carry out hearing tests such as OAE, BERA and ASSR to assess children's hearing thresholds. This examination can be an early detection if there is hearing loss in a child.

This case report was created in order to find out the appropriate initial treatment for patients with hearing loss resulting in sensorineural deafness and congenital heart disease.

2. Case Presentation

a boy aged 3 years 6 months with complaints of not responding to sounds. Patients with congenital heart disease are consulted from the Children's Polyclinic. Aloanamnesis from both parents, the patient did not respond to sounds, either from the front or from behind. Currently, patients can only babble like baba, tata, jaja without any meaningful meaning. When spoken to there is no eye contact and does not focus on the object or things given, does not turn around when called and if he wants something the patient just cries. The patient uses gadgets occasionally. The patient was able to walk at the age of 2 years.

Prenatal history: Pregnancy history The mother experienced fever accompanied by red spots on the skin at 10 weeks of pregnancy. TORCH examination (Toxoplasmosis, Rubella, Citomegalovirus, and Herpes) on mothers before and during pregnancy has never been carried out. There is no history of exposure to or contact with people suffering from measles/rubella. There is no maternal history of measles immunization, no rubella. History of other diseases such as hepatitis, hypertension, and diabetes mellitus was denied. There was no history of bleeding during pregnancy. There was no history of travel to malaria endemic areas during pregnancy. There is no history of taking medication/injections for a long period of time; there is no history of taking herbal medicine/other traditional medicines. Patients routinely have pregnancy checks every month at the midwife. The mother's nutritional history during pregnancy was adequate. Perinatal History the patient was born at term according to the gestational age, BBL 2500 grams, by caesarean section due to breech position, did not cry immediately, asphyxia (-) hyperbilirubinemia (+), no history of NICU treatment, heart disease (+). Postnatal history There was no history of high fever accompanied by red spots on the skin. History of suffering from diseases such as rubella, measles (morbilli), smallpox (varicella), mumps (parotid), jaundice and febrile seizures was denied. Children's motor development can lift their head at 8 months, sit at 12 months, and stand at 20 months and walk at 24 months.

From the physical examination of this patient, the patient was within normal limits, the local ear status examination of the ear canal was clear, there was no cerumen, no secretions, the eardrum was intact, light reflexes were present. nose and mouth examination were within normal limits. Echocardiography examination revealed congenital heart disease.

Hearing function examination in the form of OAE, BERA and ASSR as follows:

- Otoacoustic Emissions (OAE) (20/10/2023) During the DPOAE examination, it was found: - Result: refer/refer - Conclusion: There is a disturbance in the outer hair cells of the cochlea in the right and left ears

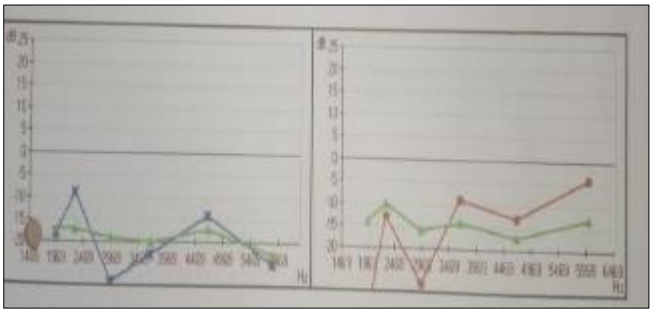

Figure 1 Examination Otoacoustic emissions (OAE)

- BERA Click examination found:

Not found V waves were at intensities up to 90 dB in both ear.

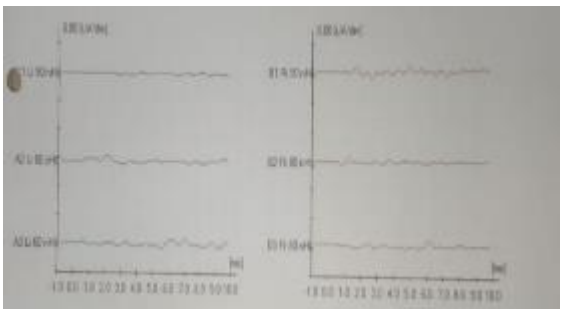

Figure 2 Examination Brain Evoked Response Auditory (BERA)

- Pada pemeriksaan Auditory Steady State Response (ASSR) didapatkan: Hasil: AD = 102,5 dB, AS = 107,5 dB Kesimpulan: Gangguan pendenganran derajat sangat berat bilateral.

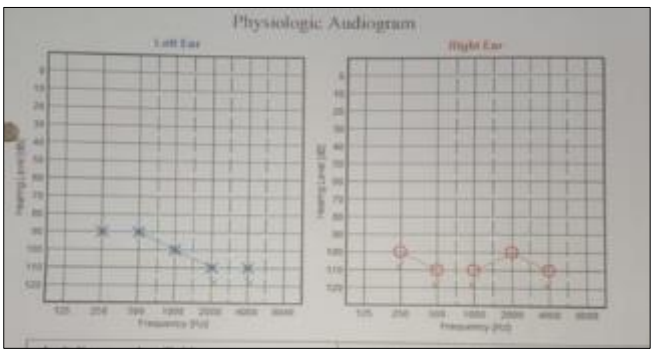

Figure 3 Examination Auditory Steady State Response (ASSR)

The patient was diagnosed with very severe sensorineural deafness in both ears + Congenital Heart Disease (CHD)

2.1. Treatment

- Installation of hearing aids
- Medical Rehabilitation Consul for speech therapy
- Continue treatment in the Pediatric cardiology department
-

2.2. Follow up and outcome

- Using hearing aids for 1 month, the patient does not feel comfortable using hearing aids, they are often removed from the ears. There is no response to sounds yet, the patient is still babbling with the words "ma-ma, ba-ba," without any meaningful meaning and there is no eye contact.
- Using hearing aids for 2 months, the patient feels comfortable using hearing aids. The patient has responded to sound, but has not yet looked for the direction of the sound. The patient is still babbling with the words "ba-ba, ta-ta, ma-ma" without any meaningful meaning and eye contact has started to occur intermittently.
- Using hearing aids for 3 months, the patient has responded to sounds and has looked for the direction of the sound. The patient is still babbling with the words "ba-ba, ta-ta, ma-ma" without any meaningful meaning and eye contact is still occasional.

3. Discussion

Patient MA, Male, aged 3 years 6 months was consulted from the Children's Polyclinic to the ENT-HNS Polyclinic of RSUDZA Banda Aceh with complaints of no response to sound. From the alloanamnesis of both parents with complaints since the age of 4 months the child did not respond when called and also did not respond to loud sounds. Until now the patient still cannot speak. Then the patient was consulted to a Pediatric Cardiologist and it was found that the child had a heart disorder.

Verbal language perception, development, and usage is strongly related to the auditory sense. Therefore, the presence of hearing loss – even to a mild extent – has a negative effect on speech–language development in hearing-impaired children, and delays acquisition of linguistic, social, academic, and sensory abilities. Further, as speech and language development are prerequisites for cognitive development, an auditory defect may affect and impair the hearing-impaired child's cognitive ability.[9,]

Patients with congenital heart disease, based on research conducted by Seppala et.al, congenital heart disease occurs in 52.2% of CRS cases. As a result of the development of heart embryogenesis along with the embryological development of the eye lens and the inner ear, so that there are abnormalities in the ear and also abnormalities in the heart. Organs affected by rubella virus infection depend on gestational age and stage of organogenesis. Cardiovascular abnormalities in CRS can be in the form of patent ductus arteriosus (PDA), atrial and ventricular septal defects and pulmonary stenosis, and various other malformation variations. Cardiovascular defects that occur in the form of tissue death in endothelial cells both in capillary blood vessels and larger blood vessels in the placenta and myocardium, impaired septal development, heart malformations especially PDA and pulmonary stenosis are related to proliferation of fibromuscular tissue and occlusion of the intima of medium-sized or larger arteries, making the lumen narrower. Some congenital heart defects that occur in fetuses from mothers infected with rubella are Atrial Septal Defect (ASD), Ventricular Septal Defect (VSD) and Patent Ductus Arteriosus (PDA). The presence of heart disorders in patients is confirmed by echocardiography results showing the presence of patent ductus arteriosus (PDA).[10,11,12,13]

During the prenatal period, precisely at 10 weeks of pregnancy, it was found that the patient's mother had a fever accompanied by red spots. Rubella infection is mostly asymptomatic when it occurs in adults. Rubella virus generally causes mild illness and even 50% of cases are not diagnosed. This can be caused by symptoms of red rash on the skin, sometimes it cannot be clearly identified whether it is caused by rubella or not. Red rash on the skin is also difficult to distinguish from other skin rash diseases because it has very similar symptoms. For example, skin rash in measles, scarlet fever and exanthema subitum.[10]

This difference occurs because the fetus is protected by the development of fetal immune responses, both humoral and cellular, and by the presence of passively transferred maternal antibodies. In maternal rubella infection, which usually occurs five to seven days after maternal inoculation, the virus spreads hematogenously across the placenta, leading to potential congenital infection of the developing fetus. In maternal rubella infection with rash, the frequency of congenital infection is more than 80% during the first 12 weeks of gestation, about 54% at 13-14 weeks, and about 25% by the end of the second trimester. Any maternal rubella infection that occurs after 16 weeks of gestation carries no risk of congenital rubella syndrome in the newborn.[12,14,15]

Motor development in this patient was only able to lift the head at the age of 8 months, sit at 12 months, stand at 20 months and walk at 24 months. Delayed motor development is not in accordance with the child's development based on milestones. Development based on milestones at 6 months of age is able to lift the head, at 8 months of age can sit without assistance, at 12 months can stand without assistance, and at 15 months can walk. Causes of motor delays can be hereditary, premature babies, low birth weight, and infections during pregnancy.[16]

Based on the current alloanamnesis, the child is 3 years and 6 months old and it was found that the child cannot speak yet. A child aged 3 years and 6 months should be able to say single words such as "mama". This shows that the child's development is lagging behind the age it should be. This condition is caused by a hearing disorder in the patient so that the patient cannot speak yet.[14]

Hearing screening options include behavioral screening techniques, evoked OAE (EOAE) or automated ABR. In neonates, a sudden and continuous response to sound can cause a Moro reflex, eye blinking or the baby waking up. The interpretation of these behavioral tests is subjective, and can only detect babies with severe deafness but cannot detect mild/moderate hearing loss or unilateral deafness. If OAE is continued with AABR in 2 stages of screening, it will provide a sensitivity of 100% and a specificity of 99%.[17]

The biological basis of 0AE is the movement of tiny outer hair cells in the cochlea, producing mechanical energy that is converted into acoustic energy in response to vibrations of the organs in the middle ear. In healthy ears, the OAE that arise can be recorded simply by inserting a probe (plug) made of sponge containing a mini microphone into the ear canal to provide acoustic stimulus and to receive the emissions produced by the cochlea. If there is a disturbance when sound is transmitted from the outer ear such as debris/cerumen, disturbances in the middle ear such as otitis media or stiffness of the tympanic membrane, then the acoustic stimulus that reaches the cochlea will be disturbed and as a result the emissions generated from the cochlea will also be reduced. According to the theory that the OAE device is designed to automatically detect the presence of emissions marked with a pass result or no emissions/reduced marked with a refer result.[17]

Auditory brainstem response (ABR) is an examination to assess the function of the VIII nerve and the auditory pathway in the brainstem. The examination is performed using surface electrodes attached to the scalp or forehead and the mastoid process or ear lobule. The principle of the ABR examination is to assess changes in electrical potential in the brain after sensory stimulation in the form of sound. The sound stimulus given through the headphone or insert probe will travel through the cochlea (wave I), cochlear nucleus (wave II), superior olivary nucleus (wave III), lateral lemniscus (wave IV), inferior colliculus (wave V) then to the auditory cortex in the temporal lobe of the brain. What is important to note are waves I, III and V. ABR requires more time and trained personnel to operate the equipment and interpret the results. ABR is not affected by debris in the external and middle ear canal but requires the baby to be in a calm state (sedation if necessary), because movement-related artifacts can occur. ABR can detect conductive and sensorineural hearing loss.[17]

Based on the examination of the patient's ASSR (Auditory State Response Report), that in the right and left ears, very severe sensorineural deafness was found with a hearing threshold of >100 dB. Based on research conducted by Madden et.al from 21 who had congenital deafness, as many as 16 patients (76%) had bilateral sensorineural deafness and 5 patients (24%) had unilateral sensorineural deafness. generally sensorineural deafness. Sensorineural deafness occurs due to damage to the inner ear.[18,19,20]

Patients with hearing loss, hearing rehabilitation is needed so that communication can run smoothly. This rehabilitation can be done using hearing aids, speech and hearing exercises and surgical techniques such as cochlear implants. Cochlear implants work by producing electrical stimulation of the auditory nerve that has undergone degeneration in the cochlear hair cells so that amplification using ABD is no longer effective. [18]

4. Conclusion

A case has been reported in a male patient, aged 3 years 6 months who was consulted from the Children's Polyclinic to the ENT-HNS Polyclinic of RSUDZA Banda Aceh with complaints of not being able to speak and not responding to sound. Anamnesis, physical examination and OAE, BERA and ASSR hearing examinations have been carried out on the patient with OAE refer/refer results, BERA results did not find wave V up to an intensity of 90 dB and ASSR results with AD: 102.5 dB and AS: 107.5 dB. Patients with very severe sensorineural hearing loss (SNHL) and congenital heart disease (CHD). Congenital deafness that occurs can interfere with speech and cognitive function. So that management must be carried out immediately in children, both by observing general conditions and installing ABD or cochlear implantation

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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