NEWBORNS HEARING SCREENING WITH OTOACOUSTIC EMISSIONS AND AUDITORY BRAINSTEM RESPONSE

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ABSTRACT

Hearing loss in newborns is a serious matter, if it is not quickly diagnosed and starts early intervention, a child will experience social, speech, language, cognitive, and academic impairments. There is a method of hearing screening in newborns, which is divided into two types, universal newborn hearing screening, and targeted newborn screening. Both of these methods use OAEs and ABR as objective examination tools. The hearing screening method varies in each country, this difference is based on the test equipment used, age, frequency, professionals involved in screening, referral procedures, funding, and coverage areas. Indonesia uses two stages of screening, while Italy, America, Nigeria, France, India, and Poland use two to five stages of screening. Hearing screening of newborns using OAEs and ABR has a sensitivity of 100% and specificity of 99,3%.

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INTRODUCTION

Hearing loss is a disability that can occur at any age. A quarter of these hearing losses can occur in newborns to toddlers. The most common cause of hearing loss in newborns is due to congenital abnormalities with a prevalence of four to six babies in 1000 live births in developing countries, whereas in developed countries the incidence is two babies out of 1000 live births.[1]

Hearing function plays an important role in intellectual and social development during childhood, if disturbed will affect children's personal and social health.[1] A child is known to experience hearing loss by his family when he is late talking compared to children his age.[3] If it is known too late, of course the obstacles to be faced will be even greater. Hearing loss or hearing loss from birth will cause impaired speech, language, cognitive, and academic development.[4]

Data from the *World Health Organization (WHO)* in 2007, showed differences in the percentage of hearing loss in East Asia. Indonesia has the smallest percentage of 4,2%, Sri Lanka 9%, Thailand 13,3%, and Nepal 16,6%. Based on these data, there are more than 100 million people suffering from hearing loss in East Asia. The cause of hearing loss, 50% genetic factors, the remaining 50% is caused by environmental factors such as diseases suffered by the mother before or after giving birth, or diseases that are not known why.[5]

Hearing screening in newborns is a program to reduce the number of hearing loss events in the world which is progressively increasing. *World health orgnization* and *The Joint Committee on Infant Hearing (JCIH)* recommend *Otoacoustic Emissions (OAEs)* and *Auditory Brainstem Response (ABR)* as an objective screening tool in the screening program hearing p there is a newborn. The results of the examination continued following the hearing screening method as an early detection of hearing loss to interventions needed for therapy.[1,6]

The purpose of this referral writing is to know the groove, a method and a comparison of hearing screening of new babies born with OAE s and ABR in Indonesia and other countries.

OVERVIEW

1. Epidemiology

Hearing a sense health survey in seven provinces of Indonesia which states that 0,1% of the population suffers from hearing loss from birth. The hearing and communication center (Hearing and Communication Center) of the ENT-Community division of Medicine Faculty, Universitas Airlangga/RSU Dr. Soetomo, Surabaya identified the visit of SNHL profound patients in 2004 children aged <3 years 191 people and children aged <5 years as many as 228 people.[7] The ENT Sub-Department of Community FKUI/RSCM conducted observations of children suffering from bilateral severe *Sensory Neural Hearing Loss (SNHL)* in 2004-2005 at the perinatology unit through the Targeted Newborn Hearing Screening program with the OAEs and *Automated Auditory Brainstem Response (AABR)* method, it was found that the highest age for patients who first came for hearing examination was 1-3 years. Observations at Sanglah Hospital Denpasar in 2005-2006 illustrate the highest percentage of hearing loss in children with delayed speech is moderate hearing loss.[7]

The prevalence of hearing loss in America, there was one to three bilateral hearing loss in every 1000 live births and two to four bilateral hearing loss in every 1000 babies with *Neonatal Intensive Care Unit (NICU)* care. Infants who undergo treatment at the NICU or have specific risk factors have an incidence of 10-20 times greater hearing loss than newborns in general (Suwento *et al.,* 2010). Screening in Nigeria there are 28 congenital ear abnormalities of 1000 births.[1]

2. Risk Factors for Hearing Loss in Newborns

The Joint Committee on Infant Hearing 2007, newborns are suspected of hearing loss if there are several risk factors that occur both in mothers who have a family history of hearing loss, infection during pregnancy by toxoplasmosis, rubella, cytomegalovirus, herpes, syphilis, B Hepatitis and *Human Immunodeficiency Virus (HIV)* infection.[1,5] Infants with congenital head and facial abnormalities involving the earlobe, ear canal, and temporal bone anomalies, hyperbilirubinemia requiring an exchange of blood transfusion, use of ototoxic drugs, meningitis, premature birth (< 37 weeks), *Appearance, Pulse, Grimace, Activity, Respiration (APGAR)* index below four in the first minute and less than six in the first five minutes, use of mechanical ventilation for more than five days, and syndromes associated with hearing loss Pendred, Usher, Waardenburg, neurofibromatosis is an indication of hearing screening before the age of 6 months.[5-7]

Sensory Neural Hearing Loss related to hypoxia in newborns is mainly related to *Persistent Pulmonary Hypertension (PPHN)* and *Extracorporeal Membrane Oxygenation (ECMO)* being risk factors for hearing loss in newborns.[8] Newborns with hyperbilirubinemia are known to cause specific changes in ABR waveforms, which disappear after returning to normal bilirubin levels through exchange transfusions. [8]

3. Hearing Screening Levels

Newborn hearing screening procedures must be performed within 72 hours before discharge from the hospital. This procedure is divided into two levels of screening namely using Transient Otoacoustic Emissions (TEOAEs), at the first level then AABR at the second level. Some countries divide into three levels of screening, where there are similarities in first and second level screening, but in the third level diagnostic BERA is added.[4,7,9,10] The Screen-Rescreen-Confirmation method should be recommended in Universal Newborn Hearing Screening (UNHS) gradually using OAE and ABR. Screening is done within the first four weeks, then repeated together with the immunization schedule in six months and starting an appropriate intervention at the age of the six-month-old child. [8]

3.1 Stage One

Stage one screening is performed on infants aged 2-3 days, using TEOAEs by providing a stimulus in the form of sequential clicking sounds with a frequency range of 1,5–4,5 kiloHertz (kHz) and a wave intensity of 70 decibels (dB) Sound Pressure Level (SPL).[6,9,10,11] Results are divided into three categories, namely infants without hearing risk factors with bilateral pass responses considered to have normal hearing and excluded from further analysis, information about progressive genetic hearing loss remains given to families. Infants without risk factors with one or both sides refer responses are advised to take the TEOAEs test again one week after discharge. In the case of a pass response, children are considered to have normal hearing, whereas, in the case of a new refer response, children are referred for stage two screening. Infants with bilateral pass responses but with family history risk factors with SNHL, pregnancy, or auditory neuropathy were referred for stage two screening.[10] In a study in Lazio, Italy, newborns concerning stage one screening were re-evaluated 15 days after the first stage examination.[10].

3.2 Stage Two

Automated Auditory Brainstem Response is used in children who do not pass the first stage of screening with at least one ear giving the results of a reference and in infants/children with high risk. The examination is based on sound intensity with click stimulation at 40 dB NHL.[9,10] The examination results are divided into, infants with normal AABR or 40dB hearing intensity with no risk factors for hearing loss. Infants with normal AABR or 40dB hearing intensity with a history of high risk or hereditary hearing loss in the family are advised to test hearing function with TEOAEs, ABR, tympanometry and acoustic signal reflexes routinely every six months in the first three years, then every 12 months in three the following year, infants with abnormal results in one ear or both ears are recommended for a diagnostic ABR examination.[9,10]

3.3 Stage Three

This screening is carried out through the stages of case history, otoscopy, TEOAEs, or DPOAE (Distortion Product Otoacoustic Emissions), ABR recording and impedance under the supervision of ENT-Head and Neck experts or audiologists. All infants who do not pass the second stage must be tested within the third month of life and must start therapy and rehabilitation at the age of six months.[10] Transient Otoacoustic Emissions are more commonly used in the NHS (Newborn Hearing Screening) method worldwide because it is easier to do, have a short testing time and are considered cheaper, either as a single check or in combination with AABR. Repeated use of TEOAEs in multi-stage screening methods increases measurement specificity and has been reported to help reduce the number of more expensive secondary-level hearing evaluation.[10]

Transient Otoacoustic Emissions can be done in shorter testing times compared to AABR, but TEOAEs cannot completely replace AABR.[6] The sensitivity and specificity of TEOAEs as a screening test for hearing loss in newborns have been widely studied in several community projects. Research in Italy found that hearing screening with TEOAEs has a specificity of 70%, in another study reporting a specificity of 84% for TEOAEs in the NICU population.[10]

4. Newborn Baby Hearing Screening Methods

Hearing loss from birth can be detected early through two kinds of programs namely, UNHS is performed on all newborns up to the age of two days or before leaving the hospital. Targeted newborn hearing screening is screening for hearing loss in infants who have risk factors, one of which cares at the NICU for more than 24 hours.[6] The selection of UNHS and Targeted newborn hearing screening is based on various factors such as, cost, logistics, infrastructure considerations, targeted reference levels and the standard level of follow-up for the screening carried out. Although the same NHS basis can be used, there are clear differences in their implementation or use in screening methods.[6]

Newborn hearing screening methods are determined to achieve the ideal screening method that is to have good sensitivity and specificity and to achieve high initial and low-cost graduation rates. This is very important to know, where the factors mentioned above can have a significant impact on the success of the NHS program. The use of hearing aids to cochlear implants gives good results in general development and language at the age of three to five years.[1,6] These differences are generally based on the test equipment used, age, frequency, professionals involved in screening, referral procedures, funding, and scope of coverage.[6]

The auditory screening method uses the OAE test, which is an objective examination used to screen for hearing in newborns. There are several types of acoustic emissions carried out, namely with TEOAEs, DPOAEs, AABR, or a combination of OAEs and AABR.[4] Retrospective studies of the UNHS program in Italian public hospitals report that hearing evaluation with a combination of OAEs,and AABR can reduce errors in undiagnosed SNHL.[9]

4.1 Hearing Screening with Otoacoustic Emissions

Otoacoustic emissions are acoustic signals from mechanical processes of the Outer Hair Cells (OHC) in the cochlea which inform the cochlear's good condition. Emissions are categorized as the presence or absence of a stimulus that can evoke an OAE response to have clinical significance.[3] Use of TEOAEs in stage one, DPOAEs and AABR in stage two in Poland have a false positive result of 82,73%, due to the presence of amniotic fluid in the middle ear and dirt in the outer ear at stage one examination.[13]

Otoacoustic emissions were introduced by Kemp in 1997 as the main standard for examining newborns. Medium-intensity clicks or the appropriate combination of two sound waves can trigger the OHC movement, then biomechanics occur from the basilar membrane to produce intracochlear energy amplification and cochlear tuning. OHC movement causes mechanical energy in the cochlea to be propagated out through the middle ear system and tympanic membrane to the ear canal. Tympanic membrane vibrations produce OAE signals, which can be measured with a microphone. Amplification produced by OHC movement in the cochlea is 50 dB, then the residual energy reaching the ear canal is 0 - 15 Db.[14]

4.1.1 Indications for checking otoacoustic emissions

The main purpose of OAE examination is to assess the state of the cochlea, specifically the function of the OHC. The results of the examination can be useful for screening hearing, especially in newborns, children or individuals with hearing loss disorders, estimating hearing sensitivity within a certain range, screening disorders of the middle ear with moderate or severe degrees, examination of functional hearing loss.[14]

4.1.2 Types of otoacoustic emissions checks

OAE devices use TEOAEs or DPOAE technology. The TEOAEs device emits one short click which covers a wide frequency range while DPOAE emits two short tones on two separate frequencies.[15,16] Targeted newborn hearing screening is used to detect baby's hearing, validate other tests, and assess cochlear OHC function. Distortion Products Otoacoustic Emissions are used to assess damage behind the cochlea, generally in patients with drug ototoxicity and damage caused by noise.[17] Spontaneous otoacoustic emissions are sounds that are emitted without acoustic stimuli. Stimulus Frequency Otoacoustic Emissions (SFOAEs) are sounds that are emitted in response to a continuous tone. Both of these tools are not used clinically.[9,17]

4.1.3 Methods of checking autoacoustic emissions

Transient Evoked Otoacoustic Emissions that are temporarily generated appear with brief acoustic stimuli such as clicks or tone bursts. Distortion Product Otoacoustic Emissions are produced by a pure tone stimulus of frequency 1 (f1) and frequency 2 (f2) to the ear at the same time. The strongest frequency occurs at the frequency determined by equation 2 (f1-f2). The cochlear frequency area that is rated by DPOAE is between these two frequencies (Figure 1).

4.1.4 Interpretation of otoacoustic emissions results

Refer results only show that hearing loss is more than 30 to 40 dB, indicating abnormal middle ear or damage from OHC. The OAE test does not further measure hearing loss or hearing threshold, the integrity of sound transmission from the vestibulocochlear nerve to the brain stem is also not assessed.[16,18] Screening of hearing at birth with OAEs is reconfirmed at the age of six months and is standardized in the UNHS program.[19]

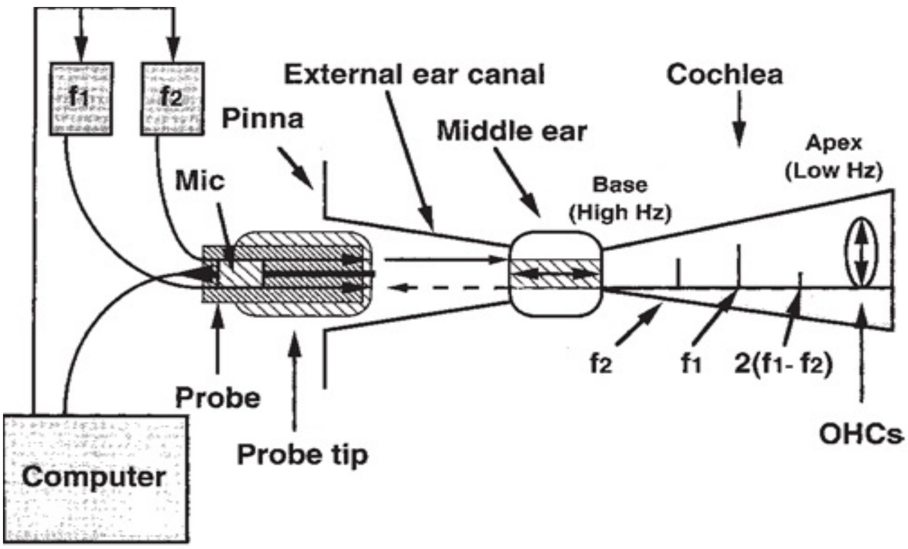

Figure 1. DPOAE examination tools and procedures.[14]

4.1.5 The advantages and disadvantages of otoacoustic emissions

Otoacoustic emissions cannot detect if there are auditory neuropathy disorders and abnormalities in the nerve so that it gives normal results, but it is different if an ABR examination is done.[18]

The examination is easy, fast and can be done in patients who are sleeping, even in a coma, because the results of the examination do not require a behavioral response. Normal otoacoustic emissions do not respond if the degree of hearing loss is above 30-40dB.[20]

4.2 Hearing Screening with Auditory Brainstem Response

Auditory brainstem response is an auditory hearing that is objective and reliable. ABR was discovered by Davis in 1976. This examination is an electrophysiological recording of responses derived from the activation of the auditory pathway to sound stimuli, ranging from the cochlea to along the brain stem, showing the integrity of nerve synchronization. This examination is noninvasive and has a high enough objective value.[14]

4.2.1 Definition of auditory brainstem response

Auditory hearing consists of an electrical response to the presence of a sound stimulus, then delivering to the peripheral auditory system, from the ear to the brain stem.[6] Auditory Brainstem Response is called the Brainstem Response Evoked Auditory (BERA), because with the right test method, this tool can record clinically at all levels of damage to the auditory nervous system, from the cochlea, auditory cortex, to responses in the brain stem. ABR waveforms show electrophysiological function as a response to sound stimuli that occur within 10ms (milliseconds) after giving a stimulus. Waves that occur within 10ms are seven waves by giving Roman numerals for each peak of the wave.[21]

Each wave shows nerve integrity in a certain area along the hearing path; wave I shows activity from the distal part of the vestibulocochlear nerve, wave II shows the activity from proximal N. VIII, wave III shows activity as high as the cochlear nucleus, wave IV shows activity from the superior olivary complex, and there may also be a contribution from the cochlear nucleus and lateral lemniscus, wave V shows the activity of the lateral lemniscus and inferior colliculus, waves VI and VII are dominated by the activity of the inferior colliculus (Figure 2).

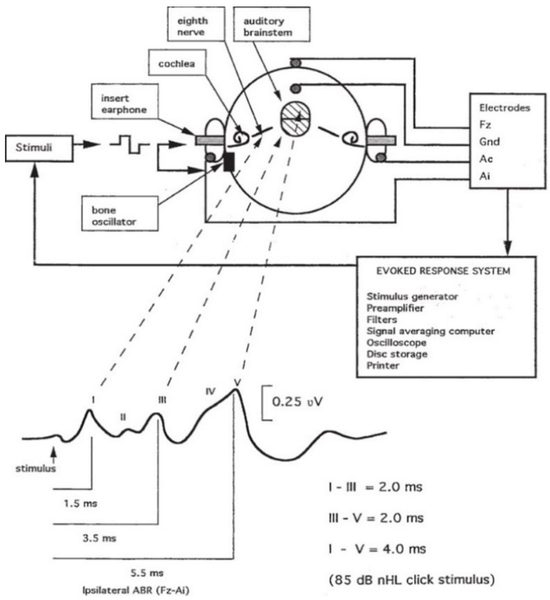

Figure 2. Inspection scheme and inspection tools in ABR.[14]

Waves IV and V sometimes combine to form complex IV-V. The amplitude component varies between subjects, but the peak latency of waves I, III, and V are relatively fixed in all subjects. V wave is the most stable wave, it is easy to be assessed even to the low-intensity sound stimulus and is clinically meaningful, therefore the assessment of predominant ABR potential is based on the latent period (time interval between the onset of the stimulus and the peak of the wave) the absolute peak.[14,21]

The application of ABR in estimating hearing sensitivity in infants and children is important in the installation of accurate and early hearing aids for babies with serious hearing loss. With the advent of the UNHS program children at risk of hearing loss can be identified early at birth.[22] Early intervention in hearing loss in infants strongly supports the use of ABD before children are 6 months old. The right hearing aid needs information about hearing sensitivity at certain frequencies. This frequency is very important for understanding speech, but it is not possible to get an estimate of hearing sensitivity at a young age with only an audiogram

4.2.2 Indications for auditory brainstem response checks

The main goal in any ABR neurodiagnostic evaluation is to record the I wave component, as a benchmark for peripheral hearing function. Subsequent interwave latency describes the function of the vestibulocochlear nerve and brain stem that is relatively unaffected by conductive or sensory hearing loss.[9]

Auditory Brainstem Response has two main uses in hearing screening. First, as an audiometric test, it provides information about the ability of the peripheral auditory system to transmit information to the auditory nerve and so on. Second, as a differential diagnosis or monitoring of central nervous system disorders. Estimated hearing threshold values are needed to find the minimum hearing threshold intensity that can produce a response on the ABR examination. Generally rely on V waves, which is the most powerful morphological aspect.[22]

4.2.3 Auditory brainstem response examination methods

ABR examination results are influenced by sedation or general anesthesia. Infants aged four months to four years old are anesthetized to minimize electrical disturbances caused by muscle activity during testing. Babies younger than four months are very likely to have an ABR examination, because in a long period after breastfeeding, the baby falls asleep.[23] The examination is canceled if the baby becomes restless and returns when the baby is calm.[16]

4.2.4 Strengths and weaknesses of the auditory brainstem response

The maximum intensity level is only around 80 dB for very short click signals and burst tones that are used to obtain ABR. Auditory brainstem responses cannot be used to estimate hearing sensitivity from severe to very severe. False-positive results may occur; a small proportion of babies with relatively normal hearing may have abnormal or absent ABR waveforms. ABR examination must be corroborated with other auditory function tests, including behavioral responses.[23]

5. Newborn screening methods in several countries

Infants and children who have high-risk factors can lose up to 50% of hearing.[24] The two most common OAE screening methods available today are TEOAEs and DPOAE.[8] The screening methods for newborn screening programs differ in each part of the world. The Indian state uses three stages of examination, TEOAEs, and AABR as the basis for the first and second stages of examination, then in stage three it is continued with AABR. The United States of America uses TEOAEs and AABR with two stages as screening.[6] The Nigerian state that uses TEOAEs in stage one gives a high referral rate, around 32,2% above the recommended value of JCIH 4%. The use of AABR in stage two reduces the level of referral results, so that it can give high results in True Negative (TN) by 60,7%, False Positive (FP) 32,7%, True Positive (TP) 5,3% and False Negative (FN) 1,3%.[6]

The auditory screening flow in France, in the first stage (T1), is performed on healthy post-natal babies in the newborn care unit at 36 hours. If the referral results of the examination can be repeated when the baby will be discharged from the primary level hospital using OAE, then proceed to stage two (T2). If at T1 refer to one side or both ears, the baby can be referred to a specialist ENT-Head Neck service center before the age of two months to be re-examined using the AABR. This is called a two-step screening. If the T2 refer to results, it can proceed to stage three. Stage three (T3) screening is carried out when the results of a referral to T1 and T2 are obtained at a special hearing center or a specialist ENT-Head and Neck service at the age of two to four weeks after birth to get re-examination up to diagnostic and management (Figure 3). In babies treated in the NICU room, hearing screening is recommended using AABR rather than OAE.[25]

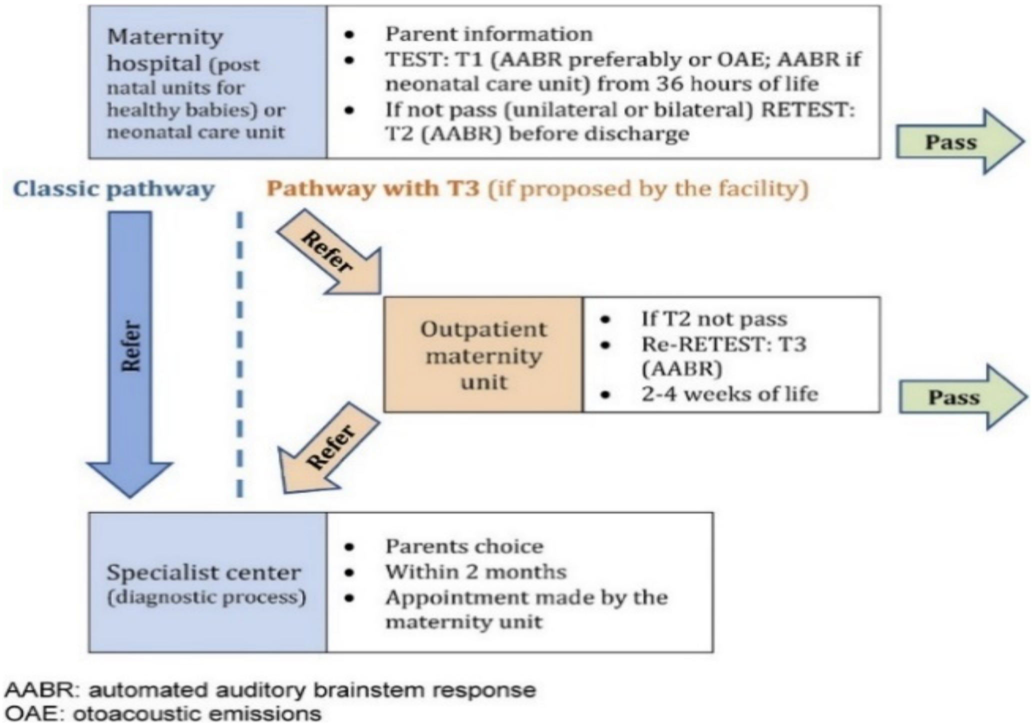

Figure 3. The screening flow and type of test used in France.[25]

Italy divides the three stages of screening based on the place/availability of facilities. Stage one screening is carried out by nurses, audiologists or neonatologists in first-level health facilities. The first stage of screening is carried out for all newborns using TEOAEs, if a pass is obtained the screening can be declared complete. On the results of a referral of either one or both ears, a re-examination is carried out before being discharged from the hospital, if the results remain refer then resuming second-level screening after 15 days.[10] Screening stage two: located in mid-level health facilities. The results of referring to newborns who have risk factors for hearing loss must be monitored with TEOAEs and ABR periodically every six months until the age of three years, then every 12 months over the age of three years for evaluation of hearing loss.[10] Stage three screening is also called the center reference. At this level all examinations are carried out, starting from repeated interviews, otoscopy, clinical TEOAEs or DPOAE, and ABR recording by an ENT-Head Neck expert until a definitive diagnosis of hearing loss is obtained and early intervention for hearing before the age of six months. This method gives a sensitivity value of 100% and a specificity of 99,3% in detecting congenital hearing loss. The error rate in the examination has a value of 0,03% and decreases at each stage of the screening (Figure 4).

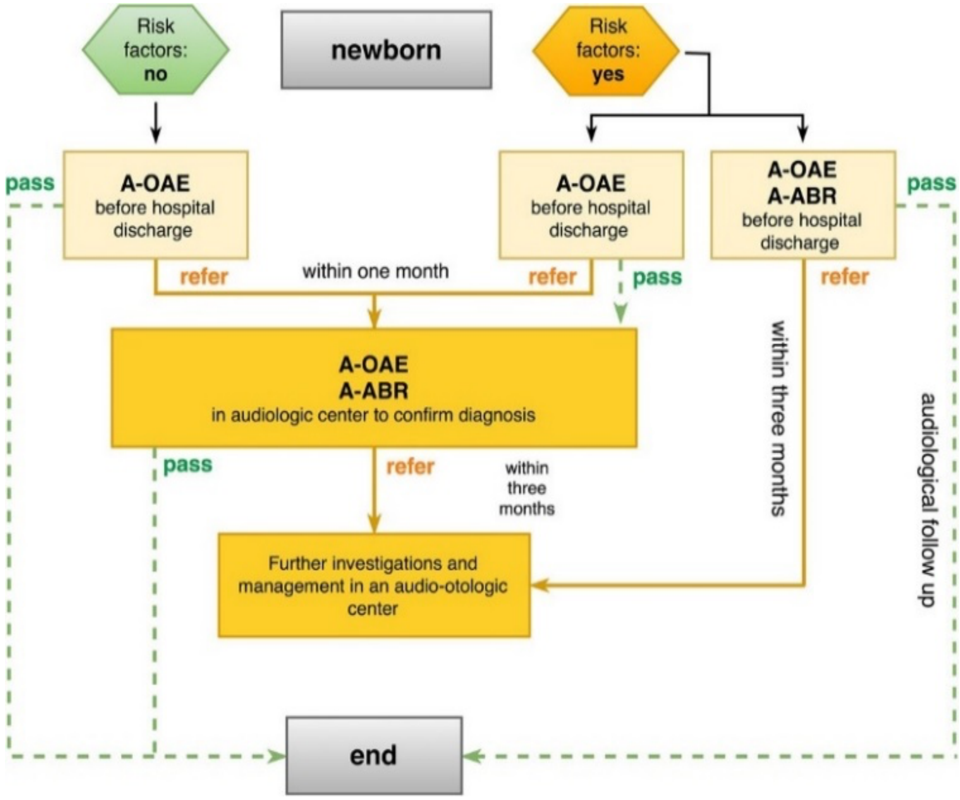

Figure 4. Screening methods and types of tests used in Italy.[10]

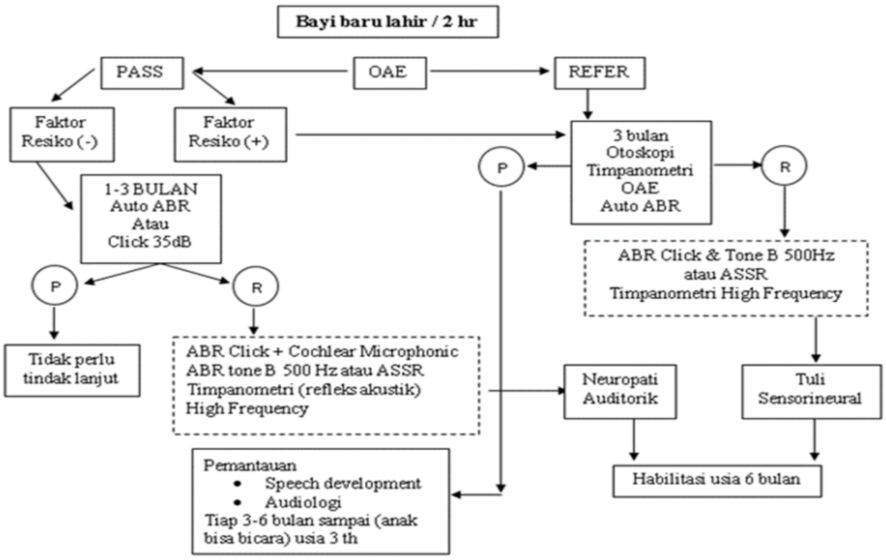

Figure 5. The flow of auditory screening in Indonesia.[7]

The method of hearing screening in newborns in Indonesia follows the path released by the Ministry of Health of the Republic of Indonesia in 2010 (Figure 5). Initial screening is done by the OAE examination after the baby is > 24 hours and before discharge from the hospital. Then at the age of three months continued with the evaluation of otoscopy, tympanometry, DPOAE, and AABR if at the first stage gave the results of the refer. If the results of stage two remain the same, a hearing evaluation is carried out with the Auditory Steady-State Response (ASSR).[7]

CONCLUSION

Hearing screening is an important program used to reduce the number of hearing loss in children. This screening uses acoustic emission devices, OAE, and ABR. Both devices were chosen because the inspection is easy to do, does not require a long time, is not invasive, and has a sensitivity and specificity of more than 90%.

Screening methods carried out in Indonesia include screening to diagnostic hearing loss, starting from the simplest to the most complex stages, but in practice not all health facilities in Indonesia have the availability of equipment or screening staff, so collaboration between the central and regional governments is needed to reduce the number hearing loss in Indonesia with intervention as early as possible.

The implementation of hearing screening in various countries varies in the number of screening stages. Tranoent Otoacoustic Emissions and AABR are widely used in screening, only a few countries such as Poland use DPOAEs. The level of accuracy of hearing screening increases when the use of OAE is combined with AABR.

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