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சுவசிரிபாய
SUWASIRIPAYA

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சுகாதார, போசணை மற்றும் சுதேச வைத்திய அமைச்சு
Ministry of Health, Nutrition & Indigenous Medicine

To all Deputy Director Generals
All Provincial Directors of Health Services
All Regional Directors of Health Services
All Directors of the Ministry of Health
Heads of all Special Campaigns
All Directors of Teaching, Provincial General and District General Hospitals
All Medical Superintendents of Base Hospitals
All Heads of Health care Institutions
All Medical Officers of Health

Newborn Screening for Congenital Deafness

Newborn Screening for Congenital Deafness is included in the 'Essential Service Package' in Sri Lanka and implemented nationally for all the newborns before discharge from the hospital after birth. About 1-2/1000 babies are born with permanent hearing loss in one or two ears. Permanent hearing loss can significantly affect the development of a child. Early detection of congenital deafness in the newborn enables a better chance of developing language, speech and communication skills.

Herewith, the guideline for Newborn Screening for Congenital Deafness is attached. Please bring the contents of this guideline to the notice of all health workers in your Province/ District/ Hospital/ Institution and take necessary steps to implement newborn screening to detect congenital deafness. Ministry of Health is in the process of procuring Oto Acoustic Emission (OAE) equipment for the hospitals without portable OAE machines. The specifications can be obtained from the Biomedical Engineering Services, Ministry of Health and Family Health Bureau to purchase OAE machines if local funds are available.

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cc:
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Guideline for Newborn Screening for Congenital Deafness

1. Introduction

A national programme has been initiated by the Ministry of Health to screen newborns for congenital deafness. One to two babies of every 1000 births are born with permanent hearing loss in one or two ears. A permanent hearing loss can significantly affect the baby's development. If deafness is corrected during early life, the child could acquire language, speech and communication skills and will lead a normal life. It will also help them to make the most of the relationships with their family or caregivers from an early age. However, if this **correction is delayed** even though the child could hear with intervention, the language ability will be significantly affected and thereby the child will be disabled.

This National programme for prevention of deafness is currently in progress and neonatal hearing screening shall be included in routine screening of the newborn. All newborns should be screened for hearing before being discharged from the hospital. The guideline issued, should be followed by the staff responsible for implementing this programme.

Test to be used in normal newborn: OAE (Oto Acoustic Emission) test

2. OAE (Oto Acoustic Emission) test

2.1 Advantages of the test

1. OAE is a noninvasive procedure, which can be performed in both ears within 5 minutes. But test time per baby may vary from 3-6 minutes dependent upon type of equipment and cooperation of the newborn.
2. This test will detect the cochlear causes of hearing loss.
3. The OAE test has a high negative predictive value, thus a good screening test.

2.2 Limitations of the test

1. Screening OAE may miss a disorder called auditory neuropathy/auditory dysynchrony.
2. Ambient noise and inexperience of the operator (technique) may affect the results.
3. External and middle ear pathologies may affect the test.

3. When to Screen the newborn for hearing

1. Testing should not be attempted prior to 12 hours of life; it is preferable to screen between 24-72 hours of life.
2. Testing should be completed as close to discharge as possible (close to 24hrs of birth). Babies born through caesarian section need to wait at least 24 hours for first attempt to allow ear canal debris to clear.
3. Screening should be conducted after a breastfeed whenever possible, to increase the chance of infant sleeping during the procedure.

4. False positive rate decreases after 12-24 hours after birth. False positive rate decreases overtime during the first 4 days of life when using OAE.
5. In newborns with risk factors for deafness (Annex 1) and newborns admitted to neonatal ICU, AABR (Automated Acoustic Brain stem Response) test should be performed.
6. In pre-term newborns, consider performing the test after 34 weeks of gestational age.
7. If the newborn is receiving antibiotic therapy, postpone hearing screening whenever possible until the antibiotic course is completed. For such newborns, hearing screening should still be performed prior to discharge. Antibiotic therapy should not be a reason for a "missed" screening.
8. Readmissions during first month of life:
A repeat hearing screening should be completed on all infants readmitted during the first month of life when there are conditions associated with potential hearing loss. eg. Hyperbilirubinemia requiring exchange transfusion or culture positive sepsis etc. Referral for an ABR should be done before discharge.

4. Who will do the test?

OAE has to be performed by a trained assigned Nursing Officer (Neonatal Hearing Screening Nursing Officer).

1. Screener/ testers should be trained by individuals with experience in newborn hearing screening techniques and practice. e.g. - Audiology Technician. Training should be competency based and involve hands-on components.
2. Screeners should also have adequate skills in soothing and calming newborns.
3. Screeners should be trained and prepared to discuss the test procedures with parents and caretakers.

5. Hearing Screening Environment:

Ideally, a space should be set aside to use for screening considering the following;

1. Close to/ in the postnatal ward
2. Has curtains
3. Has minimal noise
4. Free of electrical interference

Screening will be faster and more effective if conducted in a quiet and controlled environment.

During the time of screening

1. Turn off television/ radio
2. Ask the staff and others in the ward to be quiet
3. Minimize acoustics and electrical interference

6. How is the newborn hearing screening test (OAE) performed?

6.1 Equipment

Hearing screening of well babies will be done by a hand-held screening machine called the "Oto Acoustic Emission" or an OAE machine. This machine is an objective hearing measurement tool, utilizing ear probes that measure/ record low intensity sounds generated by the outer hair cells.

It is recommended that this machine is included in the inventory of the Nursing Officer (NO) assigned to perform the test. A trained NO from the ENT ward or any other unit (eg: Postnatal ward in a Maternity hospital) to be appointed to carry out newborn hearing screening in the hospital. Maintenance and all other procedures will be in accordance with the Department rules governing medical equipment.

6.2 Performing the OAE test

1. Inform the mother about the procedure before starting it
2. Visually inspect both ear canals for debris (wax, blood, and vernix) and clean it with rolled gauze wick if wet.
3. Change the position of the newborn, especially if the newborn has been lying on the ear you plan to test.
4. ***If both ear canals are patent and dry, OAE test will be performed in both ears.***
5. Set the ear phone probe by gently pulling the ear up and out: this will open up the canal.
6. If the probe does not pass on the first try:
Remove the probe and check for debris. Replace the tip if needed. Clean the probe if needed. Reposition the probe and repeat the screening.
7. If any abnormalities found in the ear canal, the newborn should be referred to ENT surgeon.

6.3 Informing test results to the parents

- ***If the test indicates 'pass' on both sides:*** Inform the mother "Your baby does not need any further testing at this stage".
- ***If the test fails on either side:*** "Your baby need to undergo the same test again (repeat test)"
- ***If the 2nd OAE test fails:*** "Your baby's test results warrants further testing"

6.4 Documentation and reporting of hearing

The test results should be documented in the BHT and CHDR via a 'Frank' placed in CHDR & BHT.

- Following the 1st OAE test, a form should be filled by the tester as shown in Figure 1.
 1. ***If the test passes on both sides:*** mark in front of 'Pass' box.

2. *If the test fails on either side:* mark the relevant cage and repeat the OAE test within 2-3 weeks at the same institution. If the patient cannot attend to same unit refer to local ENT surgeon.

OAE Test			
Date:			
1 → 2 →	Pass		
	Fail	B/L	R/S
		L/S	
		Date for 2 nd Test	
		
		
		(Tester's signature)	
		
		(Guardian's signature)	

Figure 1

- *If the 2nd OAE test fails:*

Refer the baby to specialized unit with AABR facility (pre identified district referral center) by completing the following form.

Newborn Hearing Screening	
Refer to:	
Name :	
.....	
Age:	Sex:
Address:	
.....	
.....	
Tel. No:	
This patient failed OAE test (B/L, R/S, L/S) twice. Please do the needful.	
.....
Date	Consultant/ MO

Figure 2

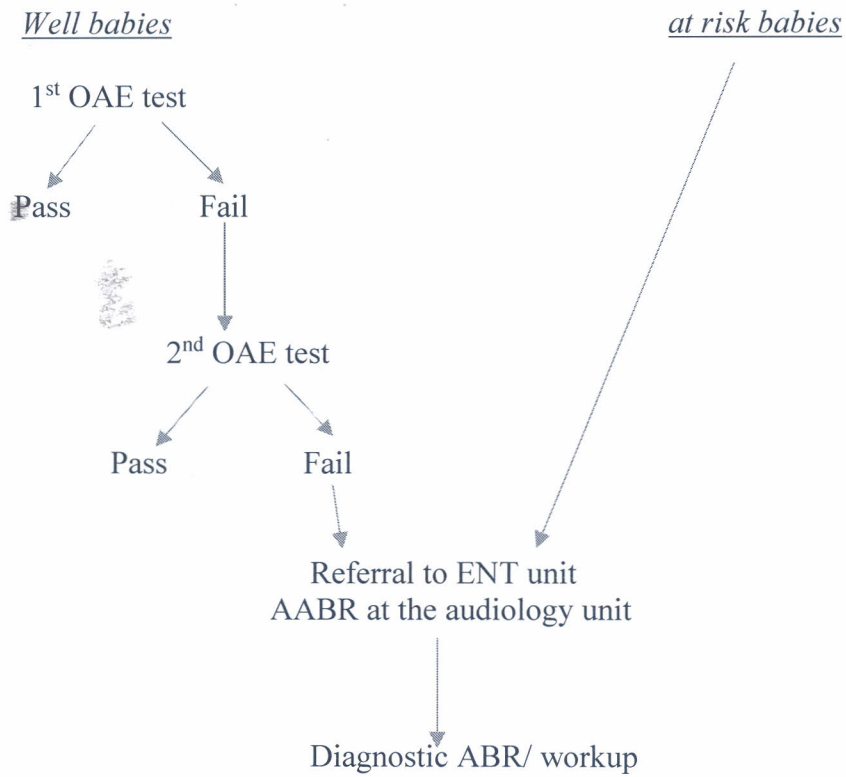
- The Medical Officer responsible for the neonatal examination in the post-natal ward should ensure the completion of the newborn hearing screening prior to discharge.
- All babies with failed screening test at discharge should be referred to ENT unit for appropriate further testing.
- The babies with failed hearing tests should be traced by the PHM / MOH of the area and refer to the relevant institution.

7. Protocol for NICU and at “risk baby” (Annex 1)

Referral note:

“Please do the hearing assessment for this patient”. Filled by MO-NICU/ Consultant Paediatrician or Neonatologist / Medical Officer (Paediatrics).

8. Protocol for Neonatal hearing screening



Annex 1 -Factors to identify 'at risk' newborns for ENT referral

Maternal factors

1. **Family history** of permanent hearing loss in childhood
2. **consanguinity**
3. **Maternal infections** during pregnancy or delivery
(Toxoplasmosis, Syphilis, HIV, Hepatitis B, Rubella, CMV, Herpes simplex, measles, mumps and others)
4. **Chemotherapy.**
Eg: Aminoglycoside gentamicin
Loop diuretics (furosemide)
Platinum based chemotherapy agents - Cisplatin.

Factors related to Newborns

1. All newborns required **neonatal intensive care for more than 7 days**
2. **Syndromes** associated with hearing loss,
Eg:
 - Waardenburg, Alport, Pendred, Jervell, Lange-Nielson
 - Neurofibromatosis, Osteopetrosis, and Usher syndrome (associated with progressive or late-onset hearing loss)
 - syndromes associated with hearing loss that include observable physical anomalies of the head, neck, and ears which frequently result in hearing loss (e.g., Down syndrome)
 - Neurofibromatosis Type II (NF2)
 - Neurodegenerative disorders Eg: Hunter syndrome
 - Sensory motor neuropathies Eg: Friedreich ataxia and Charcot-Marie-Tooth syndrome.
3. **Craniofacial & neck anomalies** including the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
4. **Culture-positive postnatal infections** associated with sensorineural hearing loss
Eg: bacterial and viral (especially herpes viruses and varicella) meningitis, Hemophilus Influenza Type B
5. **Head trauma/birth trauma**
Eg:
Basal skull (potential damage to either the cochlea or middle ear)
Temporal bone (the inner ear) fractures
Perforation of the tympanic membrane, bleeding, or disruption of the ossicular chain
6. **Ototoxic medications** given in the neonatal period
Eg: aminoglycosides (gentamycin), loop diuretics