



Audiological test and Management of hearing loss after newborn hearing screening test

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I. Introduction

- ▶ Hearing loss is the most common sensory deficit and one of the most common congenital abnormalities. It affects 432 million adults and 34 million children across the globe. It was estimated that the prevalence of bilateral moderate to severe hearing loss ranged from 1-3/1000 normal newborns and 2-4/1000 in high-risk group newborns.
- ▶ Therefore, early detection and prompt treatment are of utmost importance in preventing the unwanted sequel of hearing loss.

(<https://www.who.int/en/news-room/fact-sheets/detail/deafness-and-hearing-loss>).

- The national incidence of congenital hearing loss varies among ethnicities and diagnostic processes, and accurate statistics are difficult to obtain. The implementation of a nationwide newborn hearing loss screening program permitted the early detection of congenital hearing loss and helped determine its incidence in many developed countries.
- The incidence of congenital hearing loss in the literature is approximately 1.5 per 1,000 newborns. In detail, those figures were 1.33 and 1.86 per 1,000 newborns with various etiologies in the United Kingdom and United States.

<https://pubmed.ncbi.nlm.nih.gov/16707752/>

- In Korea, the prevalence of hearing loss was 1.2 per 1,000 newborns according to a paper published after the government-supported nationwide newborn hearing screening (NHS) pilot project .

With the advent of hearing screening protocols and the development of various auditory rehabilitation options, hearing loss is no longer an incurable disease, and many etiologies have been elucidated with the development of genetic diagnostic techniques.

If the diagnosis of hearing loss is made before 3 months of age and auditory rehabilitation is provided at the appropriate time, an infant's hearing ability and language development could be comparable to those of normal hearing children.

2. What is Newborn hearing screening ?

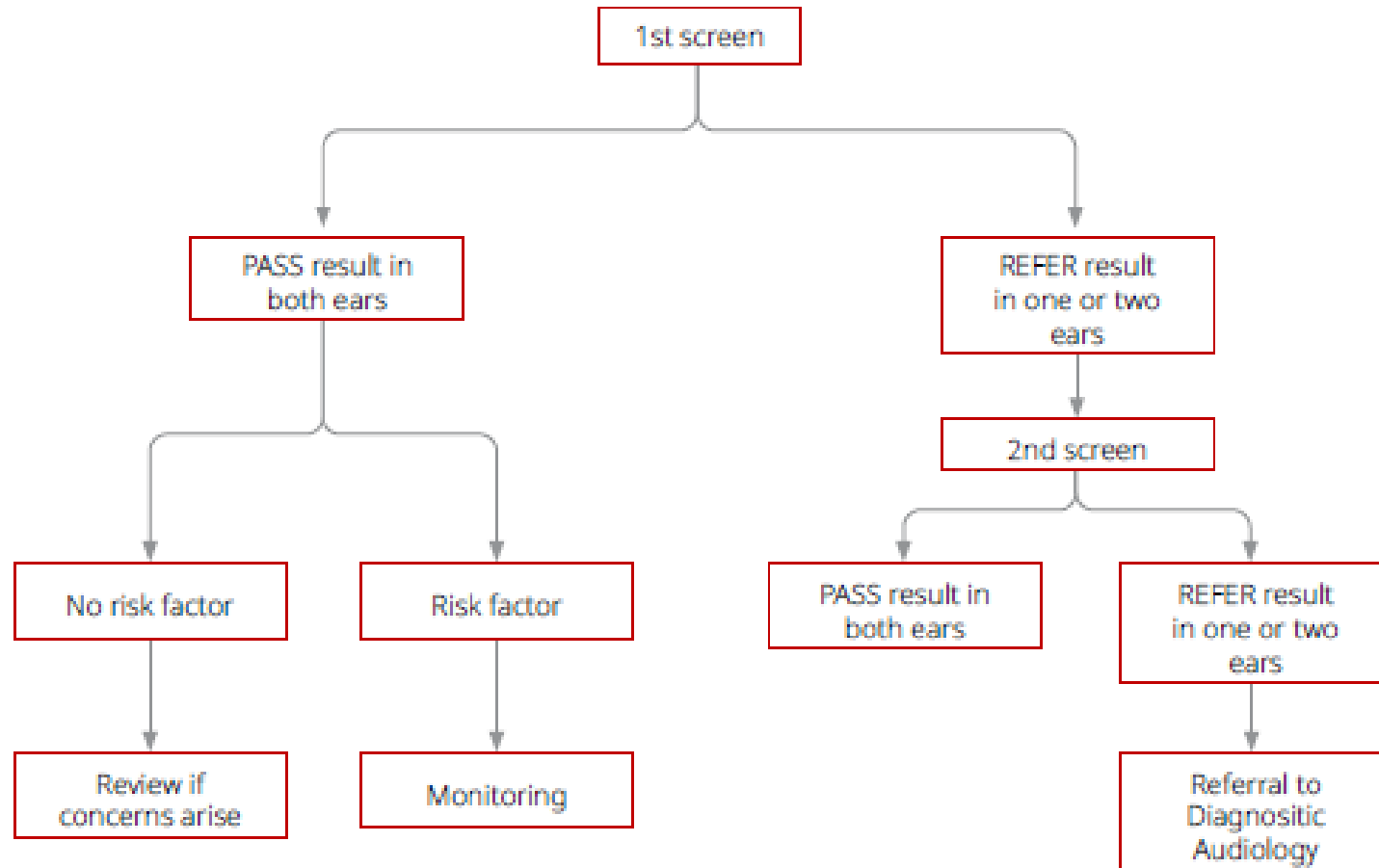
- ▶ Screening protocols for congenital hearing loss
- ▶ The concept of Universal Newborn Hearing screening for newborns was first suggested by Larry Fisch in 1957 .
- ▶ However, it was not until the end of the 20th century that it became a standard trend in almost every developed country. In Korea, NHS were introduced in obstetrics and gynecology departments in the early 2000s; in 2007–2008, an NHS project supported by the government started in some regions, and it has gradually expanded.

A. Screening protocol

A two-stage screening protocol (WHO) is important to ensure a lower rate of false positives or higher specificity of the screening programmes. This involves:

- First-stage screening, which is best performed soon after birth.
- Second-stage screening for all infants who fail the first stage screening.
- Referral for diagnostic audiology for infants who fail both the first and second stages of screening.

Two-stage screening protocol



B. GUIDING PRINCIPLES FOR NEWBORN HEARING SCREENING

Newborn hearing screening, where practicable, should be based on the “1–3–6” principle

- All infants should undergo hearing screening within the first month of life.
- All infants whose initial screening and subsequent re-screening warrant diagnostic testing, should have appropriate audiologic evaluation by no later than 3 months of age to confirm the infant’s hearing status.
- Once hearing loss has been diagnosed, the infant and family should have immediate access to early intervention service, no later than 6 months of age.



Box 1.1 The 1-3-6 principle



SCREEN BY
1 MONTH OF AGE



DIAGNOSE BY
3 MONTHS OF AGE



INTERVENE BY
6 MONTHS OF AGE

C. Age and time frame for screening

- **First-stage screening:** ideally this should take place within 1 month of age and should be conducted before the infant is discharged. First-stage screening can be performed as close to hospital discharge as possible, but there should be the possibility for a potential second-stage screening prior to discharge, if needed. When birth takes place outside of a hospital setting or where the screening programme is, for example, linked with immunization visits or well-baby clinic visits, it may not be possible to screen during the recommended 1 month period. In this case, first-stage screening should take place no later than 6 weeks of age.

- **Second-stage screening:** infants who fail the first-stage screening, should undergo a second-stage screening.
- Alternative arrangements must be made for completing the hearing screening of infants for whom it is not medically advisable or practical to do so in a timely manner, for example infants in an NICU, or on ventilators, or with severe life-threatening conditions. If screening is delayed, the procedure should be ensured once the infant

III. Hearing screening test

- ▶ As far as possible, physiological screening measures should be applied in preference
- ▶ to behavioural screening. Most developed countries currently conduct hearing screenings for all newborns.

Sensitive screening tests are commonly used and include

- automated auditory brainstem response (AABR)
- automated otoacoustic emission (AOAE)

These 2 tests is perform for newborns within 28 days of life.

The sensitivity and specificity of AOAE are reportedly 50%–100% and 13%– 91%, respectively, while those of AABR are 96% and 98%, respectively.

- First and second screening can be performed using either AABR or OAE technologies.
- With certain screening programmes, and where costs permit, AABR may be preferred;
- For infants in an NICU, AABR screening is recommended.

► Otoacoustic Emissions (OAE)

- To measure OAEs, a small probe is placed in the infant's ear canal and sound is presented by either one or two tiny loudspeakers. Any response generated by the ear is recorded with a small microphone housed inside the probe.
- Signal averaging is used to reduce the level of background noise, which comes primarily from breathing or movement.
- If an infant's cochlea is functioning normally, internally generated sounds will be recorded.
- If cochlear hearing loss exists, the cochlea either will not generate a response or it will generate a response that falls below the level that is expected from an ear with normal hearing.

- ▶ Normal external and middle ear function is important if OAE measurements are to be interpreted correctly as tests of cochlear function. Even if a cochlea is functioning normally and produces an OAE, the response may not be recorded if it is attenuated by fluid in the middle ear or any external ear canal anomalies.

► Auditory Brainstem Response (ABR)

- The normal auditory system generates small electrical currents when excited. These electrical responses are the measured response during ABR tests.
- To record an ABR, small surface electrodes are attached to the head at several places. An earphone is placed in the ear, brief sounds are played, and the electrical signals across the electrodes are recorded.

- ▶ These electrical signals, which include the ABR, are amplified and then fed to a computer, where they are averaged. Signal averaging is used to reduce the recorded level of other ongoing brain activity and to resolve the small electrical responses generated by the auditory system.
- ▶ If hearing is normal, these electrophysiological responses should be observed for low- level stimuli.
- ▶ If hearing loss exists, the threshold of these responses will be elevated.
- ▶ ABRs are less dependent on the status of the middle ear, compared to OAE tests, because the response is recorded across surface electrodes and does not have to travel back out through the middle and external ear.

Box 1.2 Considerations in selection of screening test

- Screening with OAE alone will not detect infants with auditory neuropathy, which constitutes approximately 10% of congenital hearing loss.
- The incidence of auditory neuropathy, detected by AABR, is significantly higher among infants admitted to an NICU.
- When using OAE, transient-evoked OAE (TEOAE) has greater sensitivity, as it can detect hearing levels as low as 30dBHL.
- Both OAE and AABR screening demonstrates high sensitivity and specificity, although specificity may be marginally higher with AABR.
- AABR may be more costly than OAE. However, it is to be noted that while the initial investment is higher for AABR, the follow-up costs may be greater for OAE due to higher “refer” diagnoses and false-positive rates.
- AABR is likely to take slightly longer to record than OAE.
- OAE is more sensitive to background noise levels than AABR.
- A combined OAE and AABR screening protocol has been reported as providing the best positive predictive value (PPV). However, the cost of purchasing both types of screening equipment may be prohibitive for many countries.

D. FOLLOW-UP

- ▶ **“Refer” result:** All infants who have a “refer” result after first screening should be followed up to ensure that they have a second screening. Infants who fail both first and second screenings should be referred for diagnostic testing and be followed up.
- ▶ Follow-up must be carried out systematically by a designated person to ensure that the required screening or diagnostic testing is completed. The steps in the follow-up pathway should be mapped out and a system established to facilitate family attendance at follow-up to ensure maximum compliance.

- ▶ **“Pass” result:** Parents/caregivers should be provided with information about the usual hearing and language milestones anticipated in the course of a child’s development.
- ▶ In situations where these milestones are not met, or where hearing loss is suspected, the child should undergo a hearing screening test, irrespective of previous test outcomes.
- ▶ This is important because hearing loss can develop at any time after birth, or be progressive in nature, which becomes apparent as the child grows.

3. Diagnostic assessment

All infants failing both stages of screening should undergo diagnostic audiology to confirm their hearing status by 3 months of age. Tests should include:

- **Objective assessment** of brainstem responses to sound stimulus for diagnosis of hearing loss through:
 - *Auditory brainstem response (ABR) testing* – the standard measure used to make a diagnosis of the nature and degree of hearing loss in each ear to ensure appropriate management.
 - *Auditory steady-state response (ASSR)* – this can be used in addition to ABR in order to gain frequency specific threshold estimates.

- **Tympanometry** to assess the middle ear function.
- **Acoustic reflex** to test middle ear function and integrity of auditory brainstem pathways.
- **Otoacoustic emissions** – when combined with an ABR test, this provides critical information for the differential diagnosis of auditory neuropathy spectrum disorder and sensorineural hearing loss.
- **Medical evaluation** to determine the aetiology of hearing loss.

4. Intervention

Intervention should ideally be initiated by the time an infant with hearing loss reaches 6 months of age. All countries should strive to achieve this. However, where this is not realistic immediately, countries can start with a more flexible goal to start management by 1 year of age.

Decisions regarding management should be made through a consultative family-based approach. Options for interventions include rehabilitative therapy to support the development of language skills, along with:

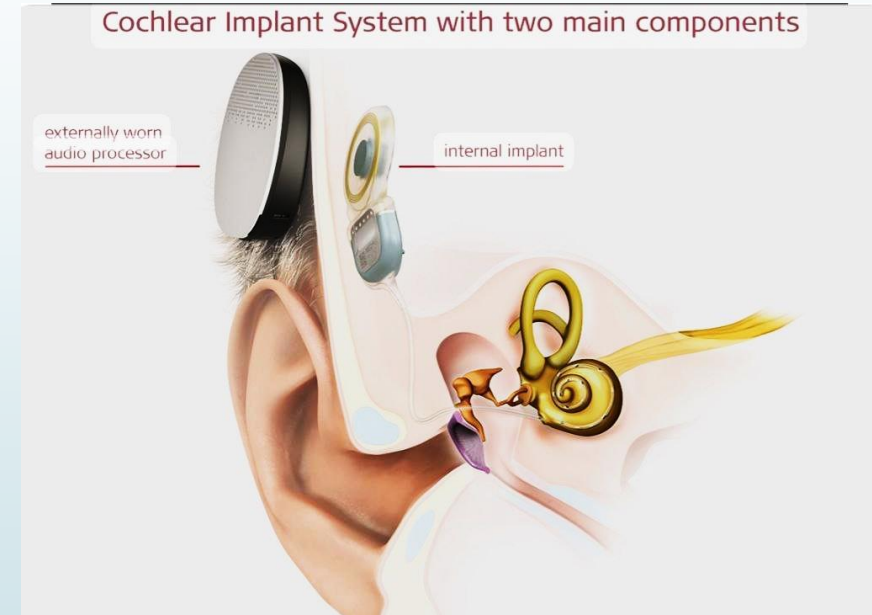
- **hearing technology use** (hearing aids or cochlear implantation);
- **sign language** learning; or
- a **combination** of the above.

In addition, parents should be directed to enrol their child in a suitable early education program.

Hearing aid



Cochlear implant

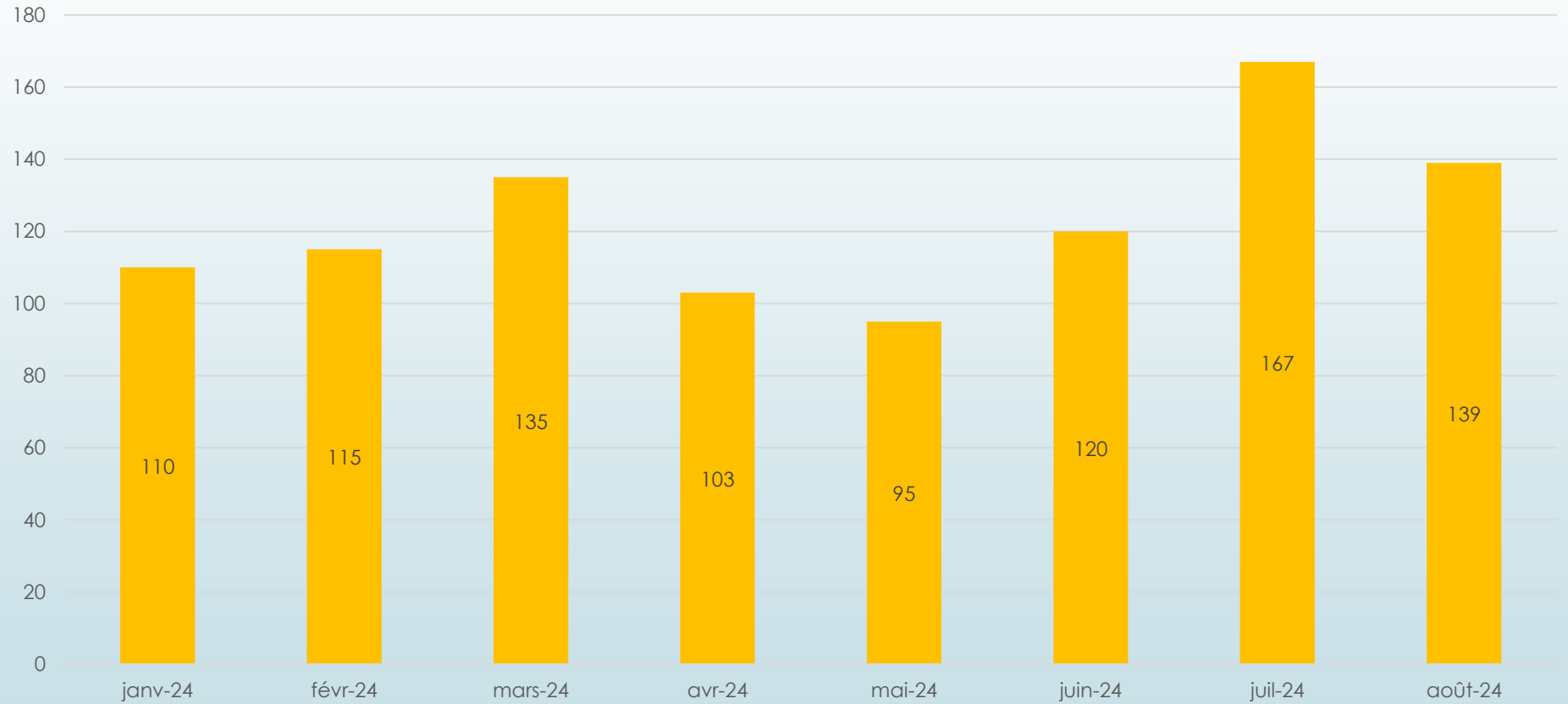


5. Newborn hearing screening in Cambodia

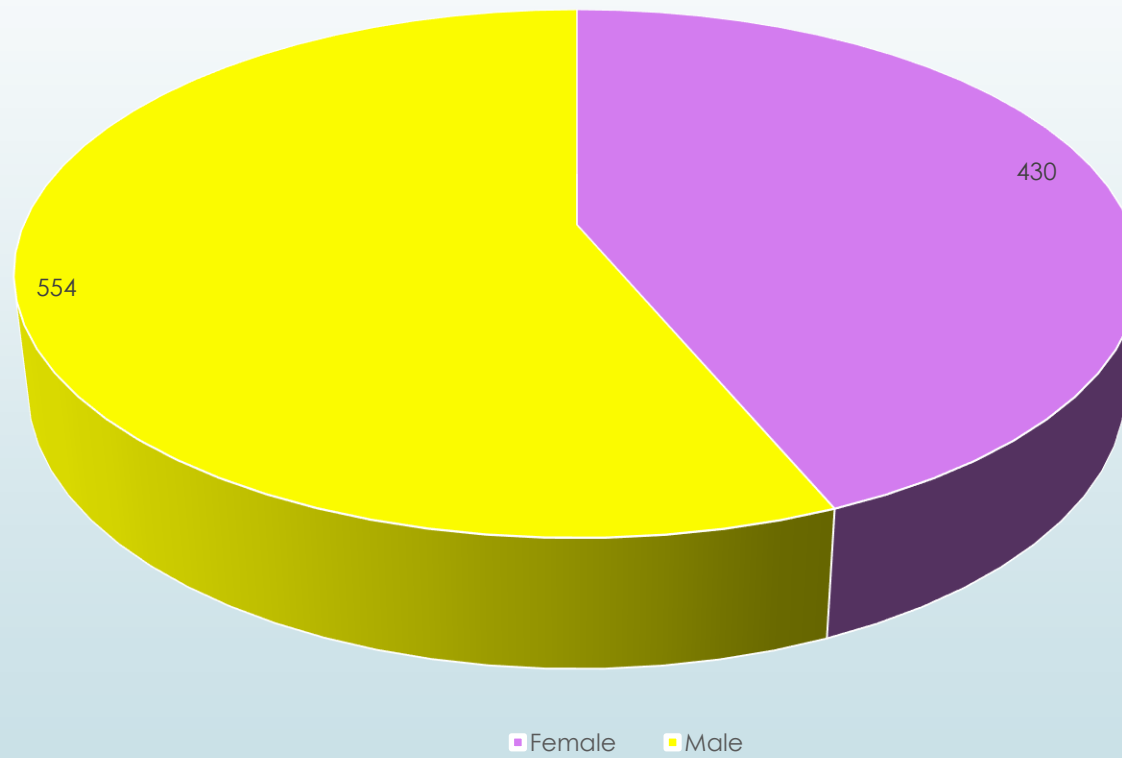
- At Preah Ang Duong hospital (national centre for ENT in Cambodia) we started in 2023 by using OAE test.
- Meanwhile, in several big private hospital in Phnom Penh has also start newborn screening in there hospital.
- In our hospital, we do the newborn hearing screening and also perform the test for newborn which refer from other hospital.

- ▶ Retrospective study on the newborn in Preah Ang Duong
- ▶ Period 8 months (from January 2024 until August 2024)
- ▶ Total 984 newborns

Number of newborn hearing screening

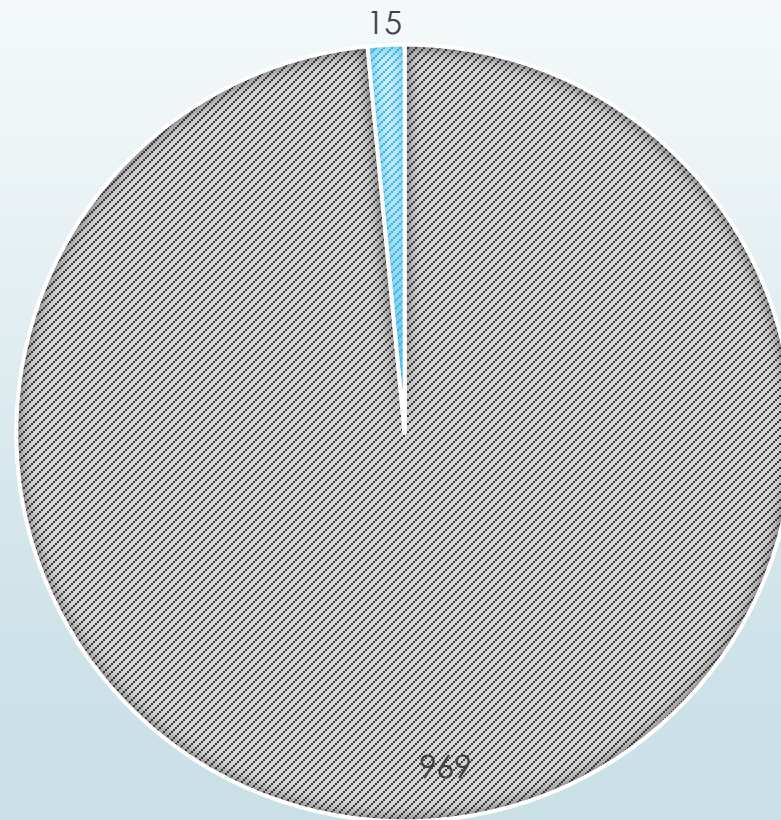


Sex of newborn



OAE RESULT

■ Pass ■ Refer



- Among the 15 newborn which test is refer, we found out that 9 of them had admit to Newborn ICU for > 48 hours due to perinatal problem such as premature, fetal distress during delivery...
- 2 newborns has a family history of hearing loss.
- 4 newborns has mother's history of viral infection during pregnancy.

6. Conclusion

- Hearing is vital for speech, language, cognition, and learning.
- Hearing loss harms the person, family, and community. Early identification and intervention of hearing loss in newborns can result in an excellent outcome.
- The rule of 1, 3, and 6 should be applied to hearing loss in newborns and infants.

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