Screening the newborn for hearing loss

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(كاهش شنوايي شايع ترين معلوليت اعصاب حسي است)

حدود **۴۷۰ میلیون نفر** در سراسر **جهان** از **کم شنوایی** رنج میبرند، که حدود **۴۰ میلیون نفر** از آنها **کودك** هستند.

دلایل اصلی رشد بالاي کم شنوایی در جهان طی سالهای اخیر:

- افزایش جمعیت **سالمندی**،
- افزایش عوامل خطر مانند ابتلا به **عفونتهاي** گوشي و دیگر عفونتها و بیماریهایي مانند سرخك، اوریون، سرخجه، منتژیت، سیتومگالوویروس،
- استفاده بیرویه و نابجا از **داروهاي** آسیب رسان به شنوایي مانند داروهاي سل و مالاریا و مصرف ناصحیح از آنتي بیوتیك هاي اتوتوکسیك آمینوگلیکوزیدي،
- قرار گرفتن افراد در معرض **اصوات بلند و آسیبهای به سر** مثل محیطهاي شغلي و نظامي و تصادفات و استفاده بي رویه از دستگاههاي صوتي شخصي در اماکن تفریحي و محل کار میباشد.

تاثير مستقيم و غير مستقيم كم شنوايي بر بسياري از جنبه های زندگي:

- كاهش توانايي افراد براي برقراري ارتباط و معاشرت،
- اختلال در تكلم
- كاهش يادگيري و ضعف در پيشرفت تحصيلی
- اختلال در كار و لذت در زندگي،
- كمك به فقر خانواده،
- انزواي اجتماعي و اختلال رفتاری و روانی
- احساس تنهايي

پايلوت طرح ملي برنامه غربالگري شنوايي نوزادان كشور: طي سالهاي ۸۳ـ۸۲

>>> اجراي برنامه غربالگري شنوايي نوزادان كشور از سال ۱۳۸۴

مجريان:

- وزارت بهداشت، درمان و آموزش پزشكي
- مركز ملي تحقيقات علوم پزشكي كشور
- مركز تحقيقات گوش، گلو، بيني و سر و گردن دانشگاه علوم پزشكي ايران

هدف:

تعيين نرخ شيوع كم شنوايي حسي عصبي شديد نوزادان كشور

محل اجرا:

۳ استان تهران، خراسان رضوي و خوزستان.

روش:

۳۵ هزار نوزاد توسط روش دو مرحله اي TEOAE در بدو تولد غربالگري شنوايي شدند .

نتايج:

نرخ **شيوع كم شنوايي حسي عصبي شديد** در نوزادان **به ازاي هر ۱۰۰۰ تولد ۱.۴ نوزاد** ميباشد.
با در نظر گرفتن **تمام حالات كم شنوايي** اين رقم به **۱۳.۷ در هر هزار تولد** برآورد شد.
بخش مراقبتهاي ويژه نوزادان **NICU** به حدود **۱۳ در هر يكصد** تولد گزارش شد

- **۳ تا ۵ درصد جمعیت کشور** داراي **درجات مختلف اختلالات شنوايي** ميباشند.

- شيوع کم شنوايي **در مراکز استانها برابر ۷/۲ در هزار تولد** زنده است.

- برآورد شده است که **سالانه ۴ تا ۵ هزار نوزاد کم شنوا** در کشور متولد ميشوند.

- ميزان اختلالات شنوايي ناشي از **مسائل ژنتيکی در ايران** نسبت به آمار جهاني بالاتر است. ۶۰ تا ۷۰ درصد مراجعين به مراکز توانبخشي کشور که دچار مشکل شنوايي بودهاند از خانوادههايي هستند که **ازدواج فاميلي** داشتهاند.

Types of hearing loss

- 1-**Conductive loss:**

caused by abnormalities of the **outer or middle ear**, which limits the amount of external sound that gains access to the inner ear (cochlea and vestibular apparatus).

transient (middle ear fluid) or

permanent (anatomical).

- 2- **Sensorineural hearing loss (SNHL):**

malfunction of **inner ear structures**, including the outer and inner hair cells of the cochlea and the eighth cranial nerve components of the auditory neural pathway.

- 3- **Auditory neuropathy (AN)**

disorder that **affects the neural processing** of auditory stimuli and may involve the eighth cranial nerve, auditory brain stem, or cerebral cortex. Screening for hearing loss using otoacoustic emissions will not detect patients with AN, as their outer hair cells function normally.

- 4- **Mixed hearing loss:**

a conductive component in combination with SNHL or AN. There is impairment in the middle ear and inner ear or auditory nerve.

Severity of hearing loss

No hearing loss – 10 to 15 dB

- Slight – **16 to 25** dB
- Mild – **26 to 40** dB
- Moderate – **41 to 55** dB
- Moderately severe – **56 to 70** dB
- Severe – **71 to 90** dB, or 61 to 80 dB (WHO)
- Profound – **>91** dB, or >80 dB (WHO)

EPIDEMIOLOGY

- **fail the screening** process = **1.6%**
- **permanent bilat. HL** subsequently diagnosed by comprehensive audiology testing = **1-3 per 1000**, making congenital HL the most common birth defect diagnosed as a result of the newborn screening process
- The prevalence of **unilateral hearing impairment above 30 decibels** (dB) has been reported as **6 per 1000 newborns**
- **Permanent hearing loss is often associated with other congenital abnormalities**, and there are **>400 syndromes** reported to be associated with permanent hearing loss

Effective SCREENING TESTS FOR neonatal hearing:

- is one that is reliable in infants **≤3 months** of age and that **detects hearing loss of ≥35 decibels** (dB) in the better ear

Two electrophysiologic techniques meet these criteria:

- Automated auditory brainstem responses (**AABR**)
- Otoacoustic emissions (**OAE**)

- **Both AABR and OAE techniques** are inexpensive, portable, reproducible, and automated. They evaluate the peripheral auditory system and the cochlea, but cannot assess activity in the highest levels of the central auditory system. These tests alone are **not suficient to diagnose hearing loss;** thus, any child who fails one of these screening tests requires further audiologic evaluation. In addition, both methods will miss mild hearing loss.

AABR:

- measures the summation of action potentials from the eighth cranial nerve (cochlear nerve) to the inferior colliculus of the midbrain in response to a click stimulus. It can detect both sensorineural hearing loss (**SNHL**) and auditory neuropathy (**AN**). Other names for this test include the screening ABR (SABR), and screening brainstem auditory evoked response (BAER). Approximately **4 percent of infants screened with AABR are referred** for further audiologic evaluation, which uses a diagnostic ABR including an evaluation by an audiologist skilled in assessing infants and young children

- AABR screening typically requires **4 to 15 minutes** for testing, although newer AABR screening equipment can complete testing in an infant in **4 to 8 minutes** in ideal conditions.

- It is important to note that an automated ABR **(AABR) is not the same as a diagnostic ABR**. AABR is a screening tool with an automated pass/fail response.

- By contrast, diagnostic **ABR** provides quantitative data (eg, waveforms) that must be interpreted by a trained audiologist, thereby determining the degree and the site of the hearing loss. As an example, delayed or absent waves suggest a neurologic or cochlear deficit.

OAE:

- testing measures the presence or absence of sound waves (ie, OAEs) generated by the cochlear outer hair cells of the inner ear in response to sound stimuli. A microphone at the external

- OAE evaluates hearing from the middle ear to the outer hair cells of the inner ear, it is used to screen for SNHL but cannot detect AN.

- OAE screening generally requires approximately **one to two minutes per ear** in ideal testing conditions.

- **Test time – OAE** tends to require less patient preparation time and a shorter test time than AABR . AABR may also present time constraints because infants need to be asleep or quiet awake when tested. In contrast, OAE can be performed when the infant is awake, feeding, or sucking on a pacifier .Response time, however, is much quicker for OAE if the infant is sleeping or quiet awake.

- **False-positive results – During the first three days of life**, there is an increased false-positive rate with OAE compared with AABR, most commonly due to transient conductive hearing loss caused by vernix occluding the external ear canal or middle ear fluid (due to amniotic fluid). . In several reports, **19 to 25 percent of newborns with abnormal OAE screening during the first three days** after birth had subsequent normal hearing in follow-up testing

Infants at risk for Auditory neuropathy (AN):

- severe hyperbilirubinemia,
- prematurity,
- perinatal asphyxia,
- craniofacial abnormalities,
- >5 d admitted to neonatal intensive care units (NICUs).

- AABR will detect the hearing loss in infants with AN, but OAE will not. Therefore screening for AN with OAE may lead to a false-negative result . Thus, **AABR** should always be used to screen hearing in infants who are at risk for AN

- **Relative costs –** Although the actual screening cost is lower for OAE compared with AABR, the overall cost of screening and audiologic evaluation may be lower with AABR because of the lower referral rate for diagnostic audiologic assessment, although this varies by location.

RISK FACTORS FOR HEARING LOSS

- Neonatal intensive care unit **(NICU) admission for ≥5 days**, especially if the neonate required mechanical ventilation or extracorporeal membrane oxygenation (ECMO) support

- **Syndromes** associated with hearing loss

neurofibromatosis, osteopetrosis, and Usher syndrome.

Waardenburg, Alport, Pendred, and Jervell and Lange-Nielsen.

- **Family history of permanent** childhood hearing loss

- **Craniofacial and ear abnormalities** (eg, cleft lip and/or palate, temporal bone abnormalities, anomalies of the pinna or ear canal)

- **Congenital infection** (particularly cytomegalovirus but also other TORCH infections such as Zika virus, toxoplasmosis, rubella, syphilis, herpes simplex virus [HSV])
- **Postnatal central nervous system infection** (eg, bacterial meningitis, HSV encephalitis)
- **Severe hyperbilirubinemia** requiring exchange transfusion
- **Perinatal asphyxia** or neonatal **encephalopathy**
- **Ototoxic mediation** (eg, aminoglycosides, diuretics)

- many of the same risk factors (**congenital infection, hyperbilirubinemia, perinatal asphyxia**) are associated with **both sensorineural hearing loss (SNHL) and auditory neuropathy (AN).**

- **craniofacial abnormalities**, are most often associated with **permanent conductive loss** due to abnormalities of the pinna, including microtia and atresia of the ear canal.

- **Selective screening** is **no longer recommended** because the available evidence suggests that this approach **misses** or delays detection of hearing loss in a significant number of patients. Thus, **universal NHS is the preferred** approach.

- A targeted screening program using the risk factors discussed above can identify only 50 to 75 percent of infants with moderate to profound bilateral hearing loss [6,45], and the time of diagnosis may be delayed [46]. In particular, infants with congenital hearing loss may not have any identifiable risk factors and selective screening will fail to identify these individuals

Goals of newborn screening

- Perform **screening** in all newborns before the age of **one month**.

- For infants who fail their screening test, perform **audiologic assessment** by **three months** of age.

- For infants found to have significant hearing loss, **start intervention** by **six months of age**. The intervention should be individualized to meet the needs of the infant and family/caregiver

With the widespread adoption of universal NHS:

- the **age at identification** of hearing loss has decreased from a range of 24 to 30 months to 2 to 3 months of age.

ویژگیهای برنامه غربالگری شنوایی موثر

- The program should have a **medical director and staff** with adequate training.

- A **minimum of 95 percent** of infants should be screened before discharge from the birth hospitalization.

Either OAE or AABR can be used for healthy term infants.

AABR should be used for infants at risk for auditory neuropathy (AN) (eg, infants admitted to the neonatal intensive care unit

- An **effective communication system** that ensures results of screening are conveyed to the family/caregiver and the primary care provider.

- A system to ensure that all infants who fail the screening test are **referred** for audiologic assessment.

- A **high follow-up rate (at least 95 percent**)

- A process for **rescreening** infants who **are readmitted within the first month** after birth for conditions associated with risk of hearing loss (eg, hyperbilirubinemia, meningitis).

APPROACH TO SCREENING DURING BIRTH HOSPITALIZATION

1- Newborn nursery

- In the newborn nursery setting (neonatal level of care 1), we suggest performing a **two-stage** rather than one-stage newborn hearing screen (NHS) primarily to reduce the number of infants with normal hearing who would be referred for further audiologic assessment

- Infants are screened initially with otoacoustic emissions (OAE), and those who fail the OAE are then screened a second time using either OAE or automated auditory brainstem responses (AABR).

- For hospitals using a one-stage NHS, we suggest screening with **AABR** since it results in a lower false-positive rate and lower referral rate for audiologic assessment compared with OAE, and it can identify infants with AN

2- Neonatal intensive care unit

- Infants cared for in a neonatal intensive care unit (NICU) for >5 days are at increased risk for hearing loss, primarily due to sensorineural hearing loss (SNHL) and AN. Thus, infants admitted to the NICU should be screened **using AABR rather than OAE** since the latter does not detect AN.

غربالگري مجدد (نوبت دوم) در نوزادان:

- نوزادان **نارس** (تكرار غربالگري كم شنوايي تا قبل از ۱ ماهگي)
- نوزادان بسيار **كم وزن** Weight Birth Low Very كمتر از ۱۵۰۰ گرم) و كم وزن Weight Birth Low كمتر از ۲۵۰۰ گرم)
- نوزادان با وزن بيش از ۴۰۰۰ گرم (نوزاد **ماكروزوم**)
- **دو و چند قلوها**
- نوزادان بستري و يا با **سابقه بستري** در بيمارستان (هر بخش از بيمارستان از جمله NICU)
- نوزادان با سابقه **دريافت و يا تعويض خون**
- نوزاداني كه **داروهاي خاص** مصرف كرده اند: مثل آمينوگليكوريدها، ديورتيك ها، ...
- نوزاداني كه نتيجه آزمون غربالگري (**نتايج آزمون اوليه**) OAEآنان مشكل دار بوده است.
- نوزاداني كه نمونه غربالگري آنان **توسط مجري غربالگري نوزادان، "ناكامل و نامناسب**" ارزيابي شده است.