

Universal newborn hearing screening

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Abstract. Hearing loss is one of the most common congenital anomalies, occurring in approximately 1-2 infants per 1000. Left undetected, hearing impairments in infants can negatively impact speech and language acquisition, academic achievement, social and emotional development. These negative impacts can be diminished and even eliminated through early intervention at or before 6 months of age. Reliable screening tests that minimize referral rates and maximize sensitivity and specificity are available. The goal of universal neonatal hearing screening is to maximize linguistic and communicative competence and literacy development for children who are hard of hearing or deaf. Audiologic and medical evaluations should be in progress before 3 months of age. Infants with confirmed hearing loss should receive intervention before 6 months of age from health care and education professionals with expertise in hearing loss and deafness in infants and young children.

Epidemiology

Worldwide estimates of the incidence of permanent bilateral sensorineural hearing loss (SNHL) of 40 dB HL or more among newborns range from 1 to 2 per 1.000.¹⁻⁵ The estimates are influenced by the level of the loss criterion: the lower the cut-off level for normal hearing, the higher will be the incidence. Its incidence is considerably higher in infants of the NICU (1-2 per 200; 1.9% bilateral and 0.6% unilateral⁶). The prevalence of hearing loss has been shown to be greater than that of most other diseases and syndromes (e.g., phenylketonuria, sickle cell disease) screened at birth. Each year, approximately 120 infants are born in Belgium with bilateral moderate-to-profound hearing loss. The bilateral to unilateral ratio was 1.10 in Flanders (personal communica-

tion). Others reported a unilateral hearing loss in 37%-48% of all hearing impaired.^{7,8}

Rationale of universal neonatal hearing screening

According to current international opinion, infants whose permanent hearing impairment is diagnosed before the age of three months and who receive appropriate and consistent early intervention at an average of 2 to 3 months after identification of hearing loss, have significantly higher levels of receptive and expressive language, personal-social development, expressive and receptive vocabulary, general development, situation comprehension, and vowel production.^{9,10} Speech development is progressively impaired by delayed age at diagnosis of hearing loss.¹¹ Impairment is measurable as early as age 3 years and has conse-

quences throughout life, leading to lower reading abilities, poorer school performance, and under- or unemployment.

Linguistic experience already alters phonetic perception in infants by six months of age.¹²

Experimental data suggest non-regressive modifications of brain organisation due to absence or inappropriate cochlear stimulation during the first half year of life.¹³ Before the advent of screening programs, the age at diagnosis of congenital hearing loss was considerably delayed. In one survey conducted before hearing screening was common, the median age at diagnosis was 13 months for infants with severe-to-profound bilateral SNHL and 17 months for those with mild-to-moderate hearing losses. Of children age 5 years with permanent hearing impairment, it is estimated that 90% have had the impairment

since the neonatal period.¹² In Flanders, the median age of diagnosis before the AABR era, was 15 months.¹⁴

According to a meta-analysis performed by Thompson *et al.*,¹⁵ universal neonatal hearing screening (UNHS) increases identification of deaf and hearing-impaired infants between 18.5% and 33% over selective screening in high-risk children (Evidence¹⁵: II). UNHS also increases the chance that diagnosis and treatment will occur before 6 months of age. (Evidence¹⁵: II).

Culbertson and Gilbert¹⁶ have also studied the effects of unilateral hearing loss on developmental abilities like mathematics, language and social abilities. They demonstrated that children with unilateral hearing loss scored considerably lower than normal (math: 30th percentile; language: 25th percentile; social: 32nd percentile). Therefore it is important that unilateral hearing loss at the screening test should also be referred for further audiological testing and follow-up. Moreover, comparison of the screening results with the definitive diagnosis has shown that in 7% of the cases a unilateral refer seem to have a bilateral moderate-to-profound hearing loss. (personal communication).

Screening methods

Even if there is a screening program, its success is dependable on its sensitivity and specificity. For decades several subjective and time consuming behavioural hearing tests such as the Ewing test were performed. In Flanders, the Ewing test was performed as a screening test in the period 1978–1998 by Kind & Gezin at

the age of 9 months.¹⁷ These tests were apparently lacking sensitivity and specificity, leading to high numbers of false negatives and false positives.

In other countries, only infants identified as being at high risk for hearing loss were routinely screened.¹⁸ According to the Joint Committee on Infant Hearing 2000 Position Statement,¹⁹ the risk factors for newborns are as follows:

- Family history of permanent childhood sensorineural hearing loss
- In utero infection such as cytomegalovirus, rubella, toxoplasmosis, or herpes
- Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal
- Neonatal indicators, specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn (PPHN) associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation (ECMO)
- Postnatal infections associated with sensorineural hearing loss, including bacterial meningitis
- Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or Eustachian tube dysfunction
- Syndromes associated with progressive hearing loss or neurodegenerative disorders
- Parental or caregiver concern regarding hearing, speech, language, and/or developmental delay
- Head trauma
- Recurrent or persistent otitis

media with effusion lasting for at least 3 months

Use of the High Risk Registry as the primary indicator for screening of newborns for hearing loss was inadequate. Screening programs in which only neonates meeting these criteria were screened, were found to exclude as many as 50% of newborns with significant congenital hearing loss. On the other hand, the presence of a high risk factor⁷ predicts hearing loss in 68%.

In UNHS programs, two types of tests are commonly used: otoacoustic emissions (OAEs) and auditory brainstem response (ABR). In comparison with the Ewing test, automated ABR (AABR: ALGO®, NATUS®) and OAEs yield far better sensitivity and specificity, are easy to use, and cost-effective.

The presence of these techniques has revolutionized the UNHS programs worldwide. In Flanders, the universal neonate screening for hearing impairment is being performed with automated ABR since mid 1998.

OAEs are highly sensitive but show lesser specificity, AABR have higher specificity. OAEs are usually easier to perform at a younger age of neonates than AABR. A drawback of OAEs is that auditory neuropathy and consequently hearing loss due to this diagnosis may be missed. Patients with auditory neuropathy have absent or severely abnormal auditory brainstem responses but have preserved cochlear outer hair cell function as shown by normal OAEs or cochlear microphonics. The prevalence of auditory neuropathy has been estimated as high as 3% to 5% of all neonatal hearing losses (1/30000 - 1/50000

of all neonates²⁰). Automated algorithms eliminate the need for individual test interpretation, reduce the effects of screener bias and errors on test outcome, and ensure test consistency across all infants, test conditions, and screening personnel.²¹

Screening strategies

To be successful a neonatal hearing screening program should endeavour to be universal, since selective screening based on high-risk criteria fails to detect at least half of all infants with congenital hearing loss. Typically, screening programs use a two-stage screening approach (either OAEs repeated twice, OAEs followed by ABR, or automated ABR repeated twice). Criteria for defining a “pass” or “fail” on the initial screening test vary, and results are sensitive to equipment, the tester’s training, the age of the neonate at the test and ongoing quality control.

In Flanders, a community-based screening program was successfully implemented by Kind & Gezin, a Federal health care agency (Decreet van de Vlaamse Gemeenschap, 29 mei 1984). Kind & Gezin coordinates the preventive care and wellness of children in Flanders and is well organised with 620 nurses working at 330 consultation bureaus over 62 Flemish regions.¹⁷

The agency’s informatic network infrastructure and manpower allows easy coordination of the services needed for screening and following-up hearing-impaired children. The ALGO[®] screening program is performed with an automated ABR (AABR) and has its main aim to identify all children with a permanent unilateral

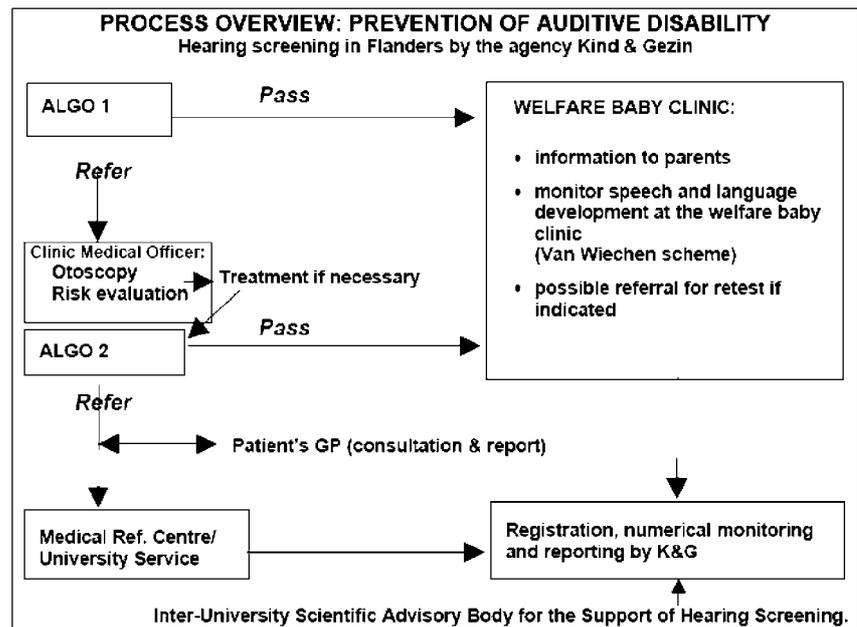


Figure 1

Overview of the procedure for audio screening in Flanders by Kind & Gezin¹⁸

as well as bilateral hearing impairment of at least 35 dB HL (Figure 1). The ALGO[®] testing is performed around the age of 4 weeks at the regional office, at home or in one of the consultation bureaus. If the infant fails at the initial test, it is repeated within 48 hours. If the test is again a uni- or bilateral referral, the baby is sent to a certified reference centre for further diagnosis and treatment. The audiologists perform an audiological test battery to include physiologic measures and developmentally appropriate behavioural techniques.

In Wallonia, an UNHS program will soon be implemented. Since ONE is not organised as its homologue in Flanders, Kind & Gezin, - ONE works with social workers instead of nurses - an alternative way for the organisation of UNHS was necessary to offer an equally qualitative screening program as in the Northern part of the country. As in most existing UNHS programs, the highest coverage and

pass rates will be sought by offering screening in hospital postnatal wards prior to discharge, with subsequent follow-up in the community. Between the 2nd and 4th postnatal day, screening with OAEs will be offered. As in most programs, a 2-stage approach will be used, in which an infant who fails the initial test is retested the day after and the infant is referred for audiologic evaluation only if he or she fails the second test or on the overall results of the 2 stages of screening.

Retesting is performed at the ENT department either in the hospital or as an outpatient within 2 weeks of discharge using OAEs and ABR.

If this auditory evaluation is pathologic uni- or bilaterally, the infant will be sent to a certified reference centre for further diagnosis and treatment.

The inherent risk of this approach is that it gives rise to a considerable percentage of lost to follow-up.²² A patient tracking

Table 1
Referral protocol: diagnostic work-up

<p>Standard procedure:</p> <ul style="list-style-type: none"> • Dysmorphic and general examination • Neurological examination • Eye examination • ECG • Urinalysis • Blood examination • CT-scan and/or MRI • DNA isolation (Connexine 26, 30) • Infection screening: Toxoplasmosis, Rubella, CMV, Herpes Simplex, Syphilis • Cytology and biochemistry <p>Complementary examinations:</p> <ul style="list-style-type: none"> • ERG , Split lamp examination • Renal echography • Cardiac echography, Holter monitoring • Rx thorax, spine... • Karyotyping • Perchlorate test • DNA isolation for Pendred and other syndromes

system with central data management is crucial to effectively manage the screening and revalidation process. In many UNHS programs, the screening to therapy coordination is the weakest point of the pathway with lost to follow-up (and treatment)¹⁵ as high as 2% to 52%.

The coverage of the screening campaign with AABR in Flanders has been around 96% since the beginning¹⁷. There are still some newborns that are missed for several reasons: moving of people, refugees, refusal of test by the parents... However, with their unique database, all lost to follow up neonates are "hunted down" in order to achieve full coverage.

A stringent cooperation protocol is needed to guarantee the most optimal follow-up. Seamless coordination between the screening program on one hand and the health and revalidation services on the other is of the utmost importance for a successful treatment strategy.

The education and training of the medical personnel performing the screening tests are equally

important and the quality must continuously be evaluated. Also communication techniques must be learned and trained to inform the parents adequately after screening.

The concepts of continual process or quality improvement to each component of the screening programs should be applied to achieve desired outcomes. Specifically, at each step in the process of care, performance mea-

asures should be undertaken to examine whether the system conforms to accepted standards of quality.

The false negative rate of UNHS programs (missed by screening) has been estimated 6%-15%. Worldwide²³⁻²⁵. False-positive rates vary among centres and depend on the strategy and timing of testing. In the Wessex study,²³ a false positive rate of the overall screening procedure (OAE followed by AABR) of 1.5% (specificity 98.5%) has been found (postnatal day 1: 1.9% to postnatal day 4: 1.1%). Kind & Gezin¹⁷ reports a false positive rate of only 0.06% (specificity: 99.94%).

If an infant has a positive result on the screening test (uni- or bilateral), the likelihood that the infant has indeed a bilateral moderate-to-profound hearing loss, is expressed by the positive predictive value (PPV: number of infants with bilateral hearing loss and a positive test divided by the total number testing positive).

In the Wessex study,²³ the overall PPV of UNHS has been estimated 6.7%. In the well-baby

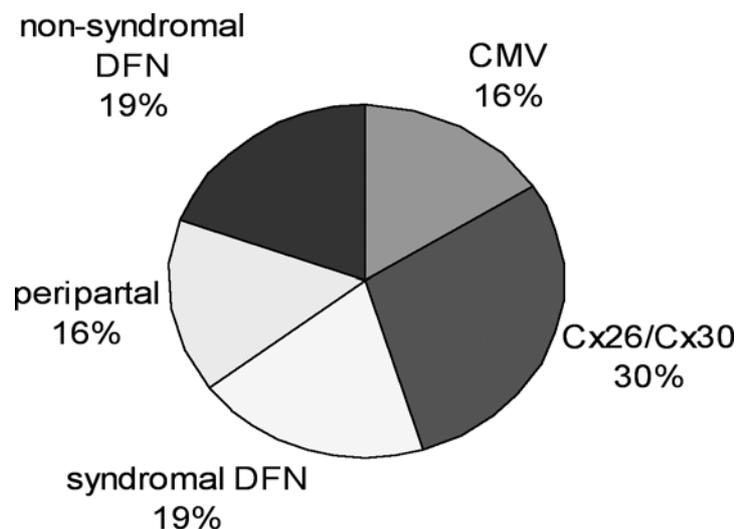


Figure 2
Diagnostic results in 100 consecutive ALGO cases: Distribution of the etiologies in infants with a diagnosis for their hearing loss

nursery, the PPV was 2.2% while for high risk-babies, the PPV was 20%. This means that for the well-baby nursery, 1 of every 45 infants referred for outpatient audiological evaluation eventually proved to have moderate-to-profound bilateral SNHL and 1 in 5 infants for the high risk-babies. In Flanders however, the screening procedure of Kind & Gezin¹⁷ in the non-NICU population, had a higher PPV of 32%.

Diagnostic workup

The gold standard for validation of the screening results is a combination of ENT and audiological consultation performed with electrophysiological testing such as diagnostic ABR, SSEP and/or behavioural testing. A test battery is required to cross-check results of both behavioral and physiologic measures.²⁶ The purpose of the audiological test battery is to assess the integrity of the auditory system, to estimate hearing sensitivity, and to identify all intervention options. Regardless of the infant's age, ear-specific estimates of type, degree, and configuration of hearing loss should be obtained. The audiologists working in a reference centre should be trained and have the technical expertise and desire to work with the infant population.

A complete case history documentation should be taken including congenital family history of hearing loss, medical factors, and risk indicators for hearing loss that may be present. Also acoustic immittance audiometry should be performed using probe tones greater than 220/226 Hz. This should also include acoustic reflexes using a higher frequency probe tone, such as 660 Hz or 800 Hz.

Visual reinforcement audiometry can be effectively employed with infants as young as 5 months. If a child has unilateral, mild, or chronic conductive hearing loss or is "at risk" for progressive or delayed onset hearing loss, ongoing audiological services should include audiological monitoring every 6 months until at least 3 years of age.

In the Wessex trial,²³ even when formal diagnostic evaluation was performed following screening evaluations, 7.4% were misdiagnosed: these infants were later found to have only a mild hearing loss. Watkin²⁷ even found a misdiagnosis in 29% of the cases. The reliability of the gold standard increases with the age at which it is performed.

In Flanders, the certified reference centres also perform a diagnostic work-up protocol for identification of the etiology of the hearing loss (Table 1). Using this diagnostic protocol in a series of 100 consecutive ALGO referrals, an etiologic diagnosis could be identified in about 45% of the cases (personal communication). A genetic cause was present in 68%, periparturient problems in 16% and congenital CMV infection in 16%. The etiologic diagnosis of the hearing loss is important for the follow-up of the infant as well for the parents if there is child wish in the future.

Conclusion

Each year, approximately 120 infants are born in Belgium with bilateral moderate-to-profound hearing loss. Universal neonatal hearing screening increases identification of deaf and hearing-impaired infants. Early diagnosis at 3 months of age and subse-

quently therapeutic intervention before 6 months from health care and education professionals maximizes linguistic and communicative competence and literacy development for children who are hard of hearing or deaf.

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CME Questions

1. Epidemiology of neonatal hearing loss: which statement is NOT true?

- A - Bilateral HL occurs 1-2 /1000 neonates/year
- B - In the NICU the incidence bilateral of HL is 1.9 %
- C - About 120 infants are born deaf in Belgium
- D - The bilateral to unilateral ratio is 1.10 in Flanders
- E - The incidence is dependent on the level of the loss criterion at screening

2. Which factor is NOT a risk factor according to the JCIH?

- A - Cytomegalovirus is a postnatal risk factor
- B - Family history
- C - Hyperbilirubinemia at a serum level requiring exchange transfusion
- D - Parental concern
- E - Mechanical ventilation

3. Regarding the rationale of UNHS: which statement is NOT true?

- A - Speech impairment is measurable at 3 years of age
- B - Phonetic perception is altered by 6 months of age
- C - Non-regressive modifications of brain organisation are measurable during first half year of live
- D - 45% of the HL at age 5 years is of neonatal origin
- E - UNHS increases HL identification between 18.5% and 33%

4. In the pre-era period of UNHS, the following statements were NOT true:

- A - The presence of a high risk factor predicts HI in 68%
- B - Using a high risk registry, 10% of newborns with hearing loss are excluded
- C - The median age of diagnosis in Flanders was 15 months
- D - Children with unilateral hearing loss scored developmentally considerably lower than normal.
- E - In Flanders, the Ewing test was used as a screening method

5. Which statement regarding the UNHS is FALSE?

- A - AABR, tympanometry and OAE are used as screening methods
- B - A two-stage screening approach is recommended
- C - 7% of unilateral refers seem to have bilateral HL at diagnostic evaluation
- D - The false negative rate worldwide is 6% to 15%
- E - Lost to follow-up and treatment in UNHS programs may be 2% to 52% worldwide

6. Which statement about auditory neuropathy is NOT true?

- A - The prevalence is between 1/30000 – 1/50000 of neonates
- B - The OAEs are always normal
- C - The ABR is always normal
- D - The cochlear outer hair cell function is preserved
- E - The cochlear inner hair cell function is preserved

7. Which statement about the UNHS in Flanders is FALSE?

- A - The UNHS is organised by Kind & Gezin
- B - The Kind & Gezin organisation has social workers as employees
- C - A two stage procedure with the AABR as screening method is used
- D - The screening is performed around 3-4 postnatal weeks
- E - The screening is done as an outpatient procedure

8. Which statement about the future UNHS in Wallonia is FALSE?

- A - The screening will be organized at the maternity ward
- B - The screening will be performed between the 2nd-4th postnatal day
- C - A two stage procedure with the AABR as screening method will be used
- D - In case of a refer, retesting will be done in the ENT department
- E - The UNHS will be coordinated by the ONE

9. Which statement about the diagnostic audiological workup is FALSE?

- A - Visual reinforcement can be employed from 5 months of age
- B - 7.4% to 29% of the diagnostic evaluations may be misdiagnosed
- C - SSEP is a new electrophysiological method to evaluate neonatal hearing loss
- D - Tympanometry with lower frequency probe tone should be used
- E - At risk children should be evaluated every 6 month till at least 3 years of age

10. Which statement about the etiologic workup is FALSE?

- A - In 45% of the cases, an etiologic factor could be found
- B - 25% of the neonatal hearing losses had a genetic cause
- C - 16% of all infants with an etiological diagnosis had a CMV infection
- D - The standard diagnostic protocol includes ECG and infection screening
- E - DNA isolation is performed to exclude Connexine 26 mutations

Answers: 1C; 2A; 3D; 4B; 5A; 6C; 7B; 8C; 9D; 10B