REVIEW ARTICLE

Early Detection of Hearing Impairment in Newborns and Infants

Martin Ptok

SUMMARY

Background: 1–2 out of 1000 newborns have markedly impaired hearing.

Methods: Review of the pertinent literature, which was retrieved with a selective search of the following databases: NHS EED (Economic Evaluation Database), HTA (Health Technology Assessment), DARE (Database of Abstracts of Reviews on Effectiveness), Clinical Trials, CDSR (Cochrane Database of Systematic Reviews), and PubMed.

Results: The current scientific evidence favors universal neonatal hearing screening (UNHS) for the early detection of hearing impairment. UNHS is best performed in two stages: first measurement of otoacoustic emissions and then automated assessment of the brainstem auditory evoked response. To be effective, UNHS programs must have a high coverage rate, high sensitivity and specificity, and proper tracking with a low rate of loss to follow-up. Children with positive screening tests for hearing impairment should undergo confirmatory testing as soon as possible and then receive the appropriate treatment. Early intervention is particularly critical for speech acquisition.

Conclusion: The early detection and treatment of hearing impairment in newborns and infants has a beneficial effect on language acquisition.

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Hearing plays a key part in learning to talk. Without speech and hearing it is difficult for interpersonal relationships to develop and thrive. In earlier times children who did not react to acoustic stimuli and were able neither to understand speech nor to acquire it spontaneously encountered severe discrimination, being dismissed as simple-minded or worse. As late as 1950, the standard policy was to wait until a hearing-impaired child had learned to speak proficiently before prescribing an individually designed hearing aid. The intention was to protect the hearing-impaired child from its own imperfect speech. Early detection of impaired hearing thus seemed superfluous, since in any case a hearing aid would not be prescribed until later (1).

Growth in understanding of the functional and morphological maturation of the auditory system and of the complexities of emotional and social development in children, coupled with advances in hearing technology, have led to a fundamental change in attitude. Today, it is axiomatic that hearing impairment should be detected and treatment initiated as early as possible. Following a ruling by the Federal Joint Committee (*Gemeinsamer Bundesausschuss*) that came into effect on 1 January 2009, all newborn children in Germany are entitled to hearing screening (2). Universal newborn hearing screening (UNHS) is either recommended or already practiced and legally regulated (nationally or regionally) (4) in many other European nations, e.g., Austria (e1), Great Britain (e2), Italy (e3), and France (e4), as well as in various countries in Asia (e5) and in the whole of the USA (3).

In deciding whether to establish a system for early detection of hearing impairment in newborns and infants, a range of questions had and have to be answered, for example:

- Will screening of newborn children always detect hearing impairment? How high are the sensitivity and specificity of UNHS? What proportion of newborns are screened? How high is the rate of children who have conspicuous findings but are lost to follow-up? Is it better to screen all newborns or only those with risk factors?
- What benefits does early detection of hearing impairment bring? Do children in whom hearing impairment is detected early by UNHS develop better than those who do not undergo UNHS and whose hearing problems are discovered later?

- What potential risks are involved in newborn hearing screening, e.g., by unnecessarily alarming parents whose children have a false-positive result?
- How soon and with what degree of certainty can confirmatory tests be carried out after a positive screening result? How quickly can treatment be initiated after confirmation of the diagnosis? Is it better to start treatment immediately or later?

Before these questions can be considered, however, two other issues have to be resolved:

- Quality: What type of hearing impairment is involved (e.g., middle ear impairment, inner ear impairment, unilateral or bilateral impairment)?
- Quantity: What is the degree of hearing impairment (e.g., expressed as hearing loss in decibels)?

This article describes how these questions can be answered or have already been answered.

Methods

A selective search of the literature (checklist at www.prisma-statement.org) was carried out in the following databases:

- NHS EED (Economic Evaluation Database)
- HTA (Health Technology Assessment) or DARE (Database of Abstracts of Reviews on Effectiveness) (http://www.crd.york.ac.uk/crdweb; last accessed on 16 August 2010)
- Clinical Trials (http://clinicaltrials.gov/ct2/search; last accessed on 17 August 2010)
- CDSR (Cochrane Database of Systematic Reviews) (http://www2.cochrane.org/reviews; last accessed on 17 August 2010)
- PubMed/Medline (last accessed on 16 August 2010)

A search on the terms "infant", "newborn", "child", "pediatric", "paediatric", "hearing loss", "hearing impairment", and "deafness" yielded 21 135 results. Restriction to the years 2007 to 2010 identified 3146 evaluable publications.

The reason for excluding earlier studies was that those before 2007 were covered in an extremely thorough survey by the Institute for Quality and Efficiency in Health Care (*Institut für Qualität und Wirtschaftlichkeit im Gesundheitswesen*, IQWiG) (5). Further restriction to publications including the terms "clinical trial" or "comparative study" narrowed the sample down to 379 studies.

Screening of the abstracts of all 379 publications was followed by full-text analysis of the studies featuring empirical data.

Prevalence

Different studies broadly agree that one or two of every 1000 newborns have a hearing impairment that on current evidence warrants treatment or observation, i.e., permanent hearing loss with a lowering of the absolute threshold of hearing for speech perception by at least 35 dB (e6–e8). To date, the UNHS study groups have concentrated on detecting such hearing impairments as early as possible (1).

Classification of hearing disorders

The term "hearing impairment" is of little practical use without further qualification. It is advisable to classify disorders of hearing according to:

- Quality and location
- Cause
- Severity.

Quality and location

The hearing process can be divided into sound conduction, transformation of sound waves into bioelectrical signals, and neural processing. A hearing disorder may involve only one or a combination of these functions. Hearing impairments can thus be classified as follows:

- Impairments of conduction (defective transport of sound waves from the external environment to the inner ear)
- Sensory impairment (defective sensation and transformation of stimuli between the base of the stapes and the first neuron of the auditory nerves)
- Retrocochlear and central hearing impairment and auditory perception disorders (defective transmission, processing, and perception of stimuli)
- Combined hearing impairments.

Particularly important in the context of early detection of hearing disorders in newborns and infants are conductive and sensory impairments, together with auditory neuropathy, which is occasionally included among the central hearing impairments.

Causes

As with other sensory impairments, there are hereditary and non-hereditary or congenital and pre-, peri-, or postnatal causes of hearing disorders. While the cause of conductive hearing loss can usually be identified relatively simply (e.g., by means of otoscopy in the case of tympanic effusion or accumulation of earwax), even thorough diagnostic investigation fails to uncover the reason for around half of the cases of inner ear hearing impairment in childhood. Approximately 50% of severe hearing impairments arising in the inner ear are thought to be hereditary in origin. The precise causes of central auditory perception disorders cannot be established.

Severity

Classification of severity is usually based on the average hearing loss in the frequency range of normal speech. Thus the hearing impairment is described solely in terms of the absolute threshold of hearing. However, the principal function of hearing is to detect rapid changes of frequency and intensity in acoustic signals above the threshold, and thus to understand speech. The conventional adult classification according to speech comprehension (speech audiometric determination of whole-word comprehension and hearing loss for numbers) is not applicable to newborns and infants. Moreover, such a classification provides only a "snapshot" of hearing ability. Intermittent hearing impairments (e.g., in children with recurring tympanic

Figure: Hearing screening with a handheld touch-screen device capable of both measurement of transitory evoked otoacoustic emissions (TOAE) and automated auditory brainstem response (AABR) audiometry

effusions) must also be considered as significant disorders. In such cases it may be beneficial to record how often episodes of hearing impairment occur over an extended period of time, e.g., a year, and then to decide how best to proceed.

Even precise classification according to the results of routine speech audiometric testing does not do justice to central hearing impairments and auditory perception disorders.

Diagnosis of hearing disorders in early childhood

Diagnosis of hearing disorders in newborns and infants is generally a two-stage process. As described above, the current standard is UNHS, followed immediately by confirmatory diagnostic evaluation as appropriate.

Universal newborn hearing screening

The various studies on UNHS have either measured otoacoustic emissions (OAE) (e9) or performed automated auditory brainstem response (AABR) audiometry, or both. In two-stage screening, OAE measurement is followed by AABR audiometry. In one very thorough analysis, Wolff et al. found that the reported sensitivity of OAE measurement varied from 50% to 100% and the specificity from 49% to 97% (6). A weakness of OAE measurement is that it does not detect fluctuating hearing impairments or those due to auditory neuropathy (e9–e12). A controlled study carried out in the UK employed two-stage screening: The estimated sensitivity (no follow-up of screening-negative children, assumption of at least a few false negatives) was 91.7% (95% confidence interval [CI] 74.2% to 97.7%), the specificity 98.5% (95% CI 98.3% to 98.7%) ([7, 8], evaluated by [6]).

From the practical clinical viewpoint, neither OAE measurement nor AABR audiometry is simple to perform. Although the equipment has become much easier to use *(Figure)*, it is advisable for the tests to be conducted by well-trained and experienced staff, and also to keep the referral rate reasonably low.

Another criterion of the quality of UNHS is the coverage rate, i.e., the proportion of newborns screened. Varying figures are reported in the literature. Green, for example, found that 95% of newborns were screened in the states of the USA in which UNHS was compulsory by law, against only 26% in the remaining states (9).

Whenever screening arouses suspicion of hearing impairment, the child concerned must undergo confirmatory diagnostic evaluation; the rate of loss to follow-up of a UNHS program should be kept as low as possible. To this end, functioning central registries must be set up (e13). These centers should ideally maintain registers of children who have not been screened and children with a conspicuous screening result. The screening center can then track these children and ensure that the required investigations are instigated. Despite the recommendations of the Federal Joint Committee, Germany is not yet completely covered by such tracking centers for UNHS.

Selective screening of newborns with specific risk factors has been discussed as an alternative to UNHS *(Box)*. These children have an up to 10 times higher risk of suffering an impairment of hearing that requires treatment (1% to 2% instead of 0.1% to 0.2%). However, around half of all hearing-impaired children exhibit no risk factors and would therefore not be among those screened. For this reason, selective screening is no longer recommended.

It is advisable for newborns with specific risk factors, e.g., those who have been treated for more than 5 days in a neonatal intensive care unit or required ventilation, to be referred for immediate AABR screening or go straight to (confirmatory) diagnostic evaluation. An American committee of experts recommends that children with risk factors *(Box)* be monitored/examined regularly for 3 years (10, 11).

UNHS is worthwhile only if (a) the diagnosis is confirmed and treatment instigated without delay and (b) these measures then have a positive impact on patient-relevant endpoints, i.e., if hearing-impaired children who are diagnosed early develop better with regard to speech acquisition than those whose hearing disorders are detected and treated later. No relevant prospective randomized trials have been conducted for ethical and moral reasons, but cohorts with and without UNHS have been compared. Sininger et al. found that final diagnosis and commencement of treatment were some 24 months earlier in hearing-impaired children who had undergone UNHS than in those who had not been screened (e14). The few studies on receptive speech skills (speech comprehension) show a significant benefit of UNHS; regarding expressive speech skills, there was at least a trend towards better development in UNHS children (for example [7, 12–14]). Wolff et al. are quite right to point out that the clinical significance of these advantages is unclear (6) (see also [e15]); however, the findings of the DECIBEL Collaborative Study Group indicate that newborn hearing screening is superior to distraction audiometry in the ninth month of life not only in terms of development of social behavior, gross motor skills, and both receptive and expressive speech skills, but also with regard to quality of life as assessed by questionnaire (15).

The potential drawbacks of UNHS include unnecessary alarming of the parents in the case of false-positive findings. However, studies have failed to confirm any such disadvantages. Since UNHS (as desired) leads—or is intended to lead—to early detection and thus to prompt treatment, the risks and drawbacks of early treatment can be counted as (late) disadvantages of UNHS. Case studies and case reports have described an increased rate of meningitis in children who receive a cochlear implant (CI) at an early age. Reefhuis et al. reported incidence of 138.2 cases per 100 000 person-years, 30 times higher than in a comparative cohort (e16). It must be pointed out, however, that most reports describe the use of a CI electrode that is now no longer employed.

BOX

Hearing impairment in early childhood: signs and risk factors (11)

- Concern on the part of parents/guardians regarding the hearing, speech development, or general development of their child
- Family history of permanent hearing impairment in childhood
- Stay of more than 5 days in the neonatal intensive care unit, possibly including the need for ventilation, extracorporeal membrane oxygenation, assisted breathing, administration of ototoxic drugs or loop diuretics, and hyperbilirubinemia requiring transfusion
- Intrauterine infections such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis
- Craniofacial anomalies, including malformation of the earlobe, auditory canal, or auricular appendages and anomalies of the auditory pit and petrosa
- External signs that may indicate a syndrome involving sensorineural hearing loss or permanent conductive hearing loss, e.g., a white forelock
- Syndromes involving immediate, progressive, or late-onset hearing loss, such as neurofibromatosis, osteopetrosis, and Usher syndrome; other complexes associated with hearing disorders are Waardenburg, Alport, Pendred, and Jervell-Lange-Nielsen syndromes
- Neurodegenerative diseases such as Hunter syndrome or sensorimotor neuropathies such as Friedreich ataxia and Charcot-Marie-Tooth syndrome
- Demonstration in culture of infections associated with sensory hearing loss, including bacterial or viral (especially herpes or varicella) meningitis
- Head injury, particularly fractures of the skull base or petrosa requiring inpatient treatment
- Chemotherapy
- Otitis media recurring frequently or persisting for more than 3 months

Although the data do not permit definitive conclusions, it is currently not thought that UNHS entails any risks that might lead to it being discredited.

Confirmatory diagnostic evaluation

If UNHS indicates that a child may have a hearing impairment, complementary investigations to confirm the diagnosis should be instigated as soon as possible. These examinations should be able to definitively confirm or exclude hearing impairment, and if a hearing disorder is present they should provide precise qualitative and quantitative characterization (e17). For the above-mentioned reasons this is practically impossible; however, confirmatory diagnostic procedures can describe hearing impairment, in the sense of permanent hearing loss of 35 dB or more, well enough for appropriate treatment to be instigated.

Pedaudiology

Hearing tests can fundamentally be divided into subjective and objective examinations. In subjective tests a sound stimulus is offered and the examiner observes and evaluates the child's reaction. Objective hearing tests are not truly objective, but are termed so because they do not require cooperation by the child. The auditory stimulus is presented (semi-)automatically, and simultaneously specific neurobiological reactions are registered (16).

Declau et al. (17) published data on confirmatory diagnostic evaluation after positive UNHS in a large group of children: Of 170 children with a positive UNHS result (corresponding to ca. 87 000 newborns screened), 5 had a tympanic effusion that regressed during the observation period. Permanent hearing impairment was confirmed in 116 children, bilateral in 68 cases and unilateral in the other 48 neonates. The average absolute threshold of hearing was 70 to 80 dB nHL. The initial (screening) AABR result was confirmed in full in 60.4% of cases. Interestingly, 11.6% of the newborns thought to have a unilateral hearing impairment on UNHS were found to have a bilateral impairment when assessed in more detail.

Objective hearing tests were used for confirmatory audiological diagnosis, but no subjective tests. This is regrettable, because even in young children subjective hearing tests yield results that help to decide how best to proceed.

Conclusion and perspective

Overall, the studies that have been carried out to date indicate that properly conducted UNHS followed by a functioning program of observation for children affected by hearing impairments yields positive results with regard to speech and general development. The risks and drawbacks of UNHS seem slight.

Nevertheless, the data on early detection of hearing impairment in newborns and infants are not very robust: Further research is clearly required. Besides the full-scale implementation of UNHS, the most important single measure for the practical realization of early detection of hearing impairments in newborns and infants in Germany seems to be the installation of a system of tracking centers covering the whole country. In the next few years we must on the one hand consider whether the rate of false-positive UNHS results can be reduced by improved assessment of the middle ear status (e18, e19). On the other hand, particularly thinking about future early interventions, we must investigate the potential utility of hybrid screening schemes, combining audiological testing with genetic screening (e.g., 35delG in GJB2/connexin mutation [e20, e21] or congenital cytomegalovirus infection [e22, e23]).

Conflict of interest statement
The author declares that no conflict of interest exists.

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KEY MESSAGES

- The current scientific evidence supports the demand for universal newborn hearing screening (UNHS) conducted by carefully trained personnel.
- A functioning system for registering and tracking both non-screened children and those with a conspicuous screening result is of crucial importance.
- Two-stage (or combined) screening yields better results than measurement of otoacoustic emissions alone; however, children exhibiting specific risk factors should immediately undergo AABR screening or (confirmatory/excluding) diagnostic evaluation.
- Conspicuous findings on screening should be swiftly followed by confirmatory diagnostic assessment and, if indicated, treatment.
- On the available evidence, the risks and disadvantages of UNHS are slight.

REVIEW ARTICLE

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