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Newborn hearing screening in the detection of hearing impairment¹

Executive Summary

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Background

In Germany, about 1 child in 1000 is born with congenital hearing impairment. Only a minority of these children are completely deaf. However, in children with hearing impairment, the maturation of neurons in the auditory system of the brain may also be insufficiently stimulated. This may lead to deficits in hearing development, which a child may compensate only with intensive interventions, or not at all. Severe loss of hearing impairs language development and may lead to lifelong damage to a child's cognitive, emotional, and psychosocial development.

Various experts have therefore called for the diagnosis of hearing impairment within the first 6 months of life in the assumption that early treatment of an affected child (e.g. with a hearing aid) can reduce the risk of such damage. In the usual routine paediatric examinations, there is often a delay in the diagnosis of hearing impairment. At the moment, the age of diagnosis for congenital hearing impairment is between 21 to 47 months of age.

In order to achieve early diagnosis and therapy, some countries, for example Great Britain and many US states, have established universal newborn hearing screening (UNHS) programmes. In these programmes, wherever possible, all newborns undergo screening with specific devices to detect signs of hearing impairment. In Germany, such screening programmes have been tested in model projects.

Aims

The aim of this report was to evaluate the benefits and harms of UNHS in the detection of hearing impairment. For such an evaluation, it is insufficient to compare only the time points of diagnosis. The focus of this report was on patient-relevant therapy goals. Through the earliest possible diagnosis and treatment of hearing impairment, developmental deficiencies in a child and their potential lifelong consequences should be avoided or at least attenuated. The effects can be measurable by means of the assessment of quality of life, hearing capacity, language development, psychosocial, emotional, cognitive, and educational development, as

well as by assessment of the adverse effects caused by false-positive or false-negative test results or unnecessary treatment.

Methods

The basis of this report was a systematic literature search for studies on 3 types of research questions. The soundest basis to answer the question as to whether universal newborn hearing screening has a benefit in children would be studies that follow the development of 2 groups of children over several years. For example, such studies would need to compare children from a region offering hearing screening with children from a region where no screening was available.

In addition, studies were also evaluated for this report in which children with early treatment were compared with children who were treated at a later stage. Such studies may also provide information on how important early treatment is. For this report, studies were also assessed that analysed the accuracy and error rate of the procedures usually applied in the early detection of children with potential hearing impairment.

In order to describe the acceptance and feasibility of UNHS programmes in Germany, as well as their main quality characteristics, reports on German model projects on UNHS were also considered in this report.

Results

Screening studies

The data from the model projects included indicate that UNHS can bring forward the time of diagnosis of congenital hearing impairment in children.

Two comparative studies investigating screening programmes in respect of patient-relevant outcomes were included in this report. With regard to language development at the ages of 3 and 8 years, both screening studies showed a tendency towards an advantage in favour of children whose hearing impairment was diagnosed in a screening programme. This may be due to earlier diagnosis in these children. Data on other patient-relevant outcomes were not available (e.g. quality of life, mental health, satisfaction, and educational and professional development). Potential harms of the screening programme (e.g. due to false-positive results) were insufficiently investigated in these studies.

Children treated earlier vs. those treated later

Four studies were included in this evaluation in which children treated earlier were compared with those treated later. Due to the study design, the studies do not allow certain conclusions. They do, however, provide indications that early treatment may be beneficial.

Studies on test accuracy

This report included 9 studies on the test accuracy of procedures applied in early diagnosis of congenital hearing impairment. Neither procedure applied, that is, the testing of spontaneous oto-acoustic emissions (S-OAE) or automated auditory brainstem response (A-ABR), has been sufficiently evaluated. One study provided information on the diagnostic quality of a screening programme in which S-OAE and A-ABR were combined. If one transferred the results of this study to 100 000 newborns, about 110 of 120 children with hearing impairment would be positively identified (sensitivity: 91.7%). The screening programme would lead to false suspicions of hearing impairment in about 1500 children; these suspicions would be dispelled after further tests (specificity: 98.5%). In reality, poor-quality screening programmes may produce substantially worse results.

Conclusions

UNHS can improve the chances that a child with congenital hearing impairment is diagnosed and treated at an earlier stage. It cannot be certainly inferred from the studies available what consequences this has for the development of these children. There are indications (not evidence) that children with hearing impairment identified by UNHS have advantages in language development. The comparison between children treated earlier vs. those treated later also provides indications that children with earlier treatment may have advantages in language development. It is insufficiently investigated how newborn hearing screening affects other outcomes relevant to the children, such as quality of life, development at school, and occupational or social situation. Programmes should therefore be designed in such a way that their quality and the consequences for the children affected can be reliably determined.

Extended executive summary

Background

The German Federal Joint Committee commissioned the Institute for Quality and Efficiency in Health Care (IQWiG) to evaluate the benefits and harms of early detection of hearing impairment in newborns by means of universal newborn hearing screening (UNHS).

Research question

The topic of this report is the evaluation of the benefits and harms of early detection of hearing impairment by means of UNHS. The focus was on patient-relevant therapy goals. Through the preferably early diagnosis and treatment of (congenital) hearing impairment, resulting developmental deficiencies in a child and their potential lifelong consequences should be prevented or at least attenuated.

A screening programme is a complex intervention whose success depends on a series of consecutive elements. The aim of UNHS is the preferably early and complete detection of children with hearing impairment who need treatment. The purpose of a screening programme depends decisively on the effectiveness of available therapies (or may also depend on other positive consequences resulting from early detection of a disorder). If children can be treated at an earlier age and the (long-term and patient-relevant) consequences of a hearing impairment can actually be verifiably reduced by bringing forward the start of treatment, then this is an indication of the benefit of such a procedure. The tests applied to diagnose hearing impairment should have sufficiently high accuracy and deliver as few incorrect results as possible.

The soundest basis for answering the question as to whether UNHS is of benefit to newborns would be studies in which the whole screening chain is examined in adequately large groups of children. The screening programme would be offered to one group, but not to the other. After an adequately long running time, the comparison can then be made to establish whether and in how many children the screening programme has prevented hearing impairment and its consequences. Such studies are complex. A preliminary search indicated that such studies of the complete screening chain had hardly been performed in newborn hearing screening. The present report therefore also examined studies which permit statements about individual screening elements (procedures for the treatment of hearing impairment and diagnostic procedures). An essential argument for the plausibility of newborn hearing screening would

be studies that show that if the diagnosis and treatment of children with hearing impairment occur at an earlier stage, this has favourable consequences. We therefore also included studies designed to compare children treated early with those treated later. Moreover, appropriate studies can compare different diagnostic techniques that might be used in hearing screening, and provide conclusions about the reliability and error-proneness of these test procedures.

If there is sufficient evidence for the benefit of early rather than late treatment and if, in addition, hearing impairment can be appropriately diagnosed in the age group of interest, this may also be seen as evidence for the effectiveness of screening.

On the basis of these considerations, objectives in 3 areas may be identified:

1. The evaluation of the effectiveness of screening programmes:

- Comparative evaluation of the benefits and harms of UNHS, versus an approach without screening, and
- Comparative evaluation of the benefits and harms of different screening strategies between each other (for example, different time points of screening, screening for different severity of hearing impairment, universal screening versus screening of children at risk)

2. Evaluation of the effectiveness of different time points of providing treatment:

- Comparative evaluation of the benefits and harms of providing treatment at different time points (early vs. later)

in each case, with regard to patient-relevant outcomes.

3. Evaluation of the quality of specific diagnostic procedures used in screening:

- Evaluation of the 2 test procedures oto-acoustic emission audiometry (OAE) and measurement of auditory evoked potentials (AEP) (e.g. by means of auditory brainstem response [ABR] testing) with regard to their diagnostic quality (e.g. sensitivity/specificity, likelihood ratios) and positive predictive values.
- Comparative evaluation of the suitability of these 2 relevant test procedures in a screening setting (e.g., time needed, influence of investigator/setting, consequences of different test quality criteria).

Methods

For the areas “screening” and “treatment”, the evaluation was performed on the basis of data from randomised controlled trials (RCTs). As a preliminary search showed that RCTs had not previously been performed to study the benefit of newborn hearing screening, for screening and therapy studies, non-randomised intervention studies and cohort studies were also considered. The outcomes selected were parameters that enabled an assessment of patient-relevant therapy goals such as quality of life, hearing capacity, language development, as well as psychosocial, emotional, cognitive and educational development. Adverse effects caused by false positive or false negative test results or by treatment were also assessed.

To investigate the accuracy and suitability of diagnostic tests, diagnostic studies were to be considered in the situation of application in newborns with unknown disease status under everyday conditions. If such studies were not available in sufficient number and/or quality, studies in newborns with known disease status were also to be considered.

In addition to the diagnostic accuracy, the outcomes investigated were also parameters that allowed statements on the suitability of the relevant procedures in a screening setting, for example time invested and the impact of the test conditions on the diagnostic accuracy.

In order to describe the acceptance and feasibility of UNHS programmes in Germany, as well as their main quality characteristics, reports on German model projects on UNHS were also considered in this report.

The systematic literature search was performed in the 11 databases MEDLINE, EMBASE, CINAHL, PsycINFO, PSYINDEX, ERIC, and the databases of the Cochrane Library (Clinical Trials [for primary publications], Systematic Reviews [CDSR], Other Reviews, Economic Evaluations, and Technology Assessments).

The literature screening was performed by 2 reviewers independently of one another.

After an evaluation of the quality of the relevant studies to be included in the report (also performed by 2 reviewers independently of one another), the results of the single studies for each separate area were collated according to therapy goals.

IQWiG’s preliminary evaluation, the preliminary report, was published on the Internet (www.iqwig.de). Interested parties could submit written comments. All written comments

fulfilling formal criteria were discussed in a scientific debate before production of the final report.

Results

A total of 2 screening studies including 120 and 50 children with hearing impairment were identified by the various steps of the literature search and included in the evaluation. One study prospectively compared alternating screening periods (with and without UNHS) (Subpopulation I), and also compared hospitals with UNHS vs. those without UNHS programmes (Subpopulation II). The other study retrospectively investigated children with hearing impairment who had either been born in hospitals with or without UNHS. Both studies showed major deficiencies regarding study and publication quality.

A total of 18 therapy studies were identified; after extracting the relevant data, 4 of these studies were included in the evaluation. The number of children who fulfilled the inclusion criteria varied between 86 and 153 in these studies, which directly compared the benefit of early compared with later treatment. All 4 studies were retrospective cohort studies; one study was population-based. Three studies showed major, and one study showed minor quality deficiencies.

A total of 12 diagnostic studies were identified, of which 9 were included in the actual evaluation. One study assessed 25 609 newborns who were initially screened in a 2-step screening programme, starting with the testing for transient evoked otoacoustic emissions (TEOAE), followed, if this test was failed, by automated auditory brainstem response (A-ABR) testing. Eight studies compared OAE with A-ABR and included 105 to 500 children. All studies showed major quality deficiencies.

The data from the model projects considered indicate that by performing UNHS, congenital hearing impairment in children can be diagnosed earlier. The 2 screening studies identified, which compared screening programmes in respect of patient-relevant outcomes, tend to indicate that children with hearing impairment identified by screening are at an advantage with regard to language development at an (average) age of 3 and 8 years compared with children whose hearing impairment was identified outside a specific screening programme or in a screening programme performed at a later age. The chances of normal speech development appear to be higher for screened children, possibly due to earlier diagnostic clarification in these children. Data on other potential long-term patient-relevant outcomes

were not available (e.g. quality of life, mental health, satisfaction, educational and professional development). Likewise, no reliable conclusion was possible on potential adverse effects of screening, as the available data were inadequate.

The 4 therapy studies included compared children given early treatment with a hearing aid or a cochlear implant with children given late(r) treatment. These studies also provided indications that early treatment may be of advantage.

The test procedures S-OAE and A-ABR used in UNHS have not been evaluated in adequately large samples of the UNHS-relevant target group – mainly healthy newborns. Only one study on the diagnostic accuracy of 2-stage screening could be identified. The results indicate that the specificity is relatively high (98.5%), with somewhat lower sensitivity (91.7%).

If the group of children (about 17%) is included who remained unscreened (even though a screening programme was offered), the sensitivity of the screening programme drops to 71.0% (95% confidence interval: 52%-86%). This means that approximately 3 of 10 children with profound hearing impairment were not identified by the screening programme. The other diagnostic studies included only allow a statement on the accuracy of measurement of otoacoustic emissions compared with the evaluation of auditory brain stem potentials. The diagnostic accuracy of the OAE varied greatly between the studies; these data do not allow a reliable conclusion.

The 6 additional reports on German UNHS model projects showed that UNHS is widely accepted in Germany too, as is evident from the very low rate of parents who refused to allow their children to participate in the screening programme. The organisational preconditions have in principle already been met. Implementation nevertheless sometimes turned out to be difficult, as seen in the comparatively low coverage rates (relative to all births in a region) and/or the high rates of children lost to follow-up. It is absolutely essential that those children initially identified as having abnormal test results in the (primary) screen must be properly tracked, which requires considerable effort.

Conclusion

There are indications that children with hearing impairment identified in UNHS programmes have advantages with respect to language development. Other patient-relevant outcomes, such as social aspects, quality of life, educational development and finally, professional situation, have not been adequately investigated for evaluation.

If the Federal Joint Committee decides to introduce UNHS in Germany, it is recommended that suitable concomitant measures should be implemented at the same time to provide quality assurance. These measures should comprise the following factors: unambiguous case definitions; specification of clear quality standards (minimal coverage rate, maximal rate of positive tests in the first step, time of confirmatory diagnostic procedures, time of the start of provision of treatment); as complete a tracking as possible of children with abnormal test results and children diagnosed with congenital hearing impairment; and identification of all children with congenital hearing impairment (including those from periods or regions without screening) at a suitable point later in time.

Key words

Congenital hearing impairment, cochlear implant, brainstem response audiometry, hearing aid, early intervention to promote hearing and language development, otoacoustic emissions, universal newborn hearing screening, systematic review