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Universal Neonatal Hearing Screening Is it obligatory, voluntary or not really necessary?

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Dr. Dr. h.c. Monika Lehnhardt

Table of Contents

Facts, Figures and Findings

Historical Highlights

Overview of the Worldwide Situation

Definition and Quality of Screening

NHS is the First Step Only

Conclusion



Dr. Dr. h.c. Monika Lehnhardt

Facts, Figures and Findings

Statistics worldwide agree that hearing impairment is the most frequent disorder in newborns. One to two out of 1.000 newborns are severely hearing-impaired or deaf, and another 2-4 suffer from moderate hearing loss. This means that we have approximately 5.000 deaf-born babies in the European Community every year, and another 5.000-10.000 have a hearing loss that requires intervention. In Germany (which is the country with the lowest birth rate in Europe) this means 600 – 1.200 deaf-born children, in Poland – 400 to 800, and in Armenia, assuming the same incidence rate, this would mean 40-80 deaf babies every year.

Experts worldwide agree: deafness does not only mean “not hearing sounds”.

Impairment of the auditory system may or will have dramatic effects on the anatomical, physiological and psychological maturation, and consequently on the development of the personality of the child.

A deaf child will normally not acquire speech and spoken language. Normal language, however, is essential for the child to achieve a proper level of intellectual, emotional and social development. And this is the basis for the child to be integrated into the world of the hearing society, to find her/his way into inclusion programmes, to achieve a higher level of education, and later in life to have better chances for employment in the adequate and desired position. This is the basis for enhanced self-esteem and self-realization, and thus for a better quality of life.

Deafness is not visible and this is one reason why in the absence of Universal Neonatal Hearing Screening hearing loss is recognized when the child is already 2 to 3 years old. At the same time, we know that the first year of life is most important for language development.

We should bear in mind what happens during the first couple of months after birth.

Between 0 and 3 months the baby

- is irritated by sudden loud noises;
- recognizes and calms to mother's voice.

Between 3 and 6 months the baby

- begins to turn eyes or head to sounds;
- stops crying when spoken to;
- begins making vowel sounds.

Between 6 and 9 months she/he

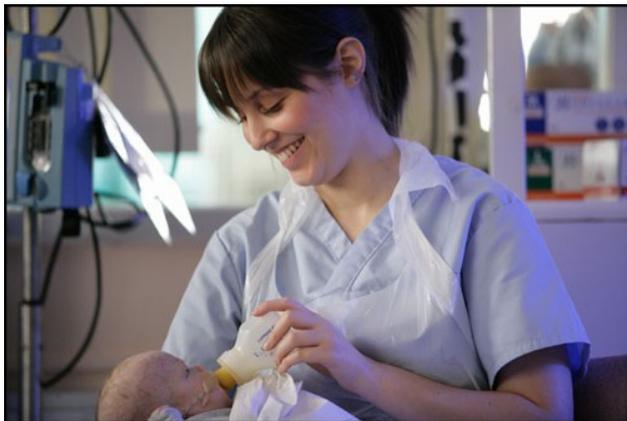


- looks for family members when her/his name is called;
- normally turns head towards sound;
- will respond to her/his name;
- responds to simple requests with gestures;
- uses voice to get your attention.

Between 9 and 12 months the baby

- understands “No”, “Bye, bye”;
- responds to soft sounds;
- looks at pictures when named;
- repeats simple sounds that we make;
- uses voice when playing alone.

A deaf baby whose severe hearing loss or deafness have not been identified and who does not get an adequate level of attention and support from their parents, will not show many of these reactions.



“The earlier the better” is a hypothesis that has been proven right over the last 15 years (see for example early and recent research by Yoshinaga-Itano, Anu Sharma, Paul Govaerts, Maree Doble, Anne Geers, Manuel Manrique, Svirsky, John Niparko, the Nottingham group with Sue Archbold and Dimity Dornan. Also see “Overview of the Evidence” presentation by Monika

Lehnhardt in Venice, Cernobbio and Vienna).

Carol Flexer makes her point by saying: *“Identification of newborn hearing loss should be considered a neuro-developmental emergency”*.

Research all over the world has shown that early intervention significantly improves receptive and expressive language skills, so there is clearly a need for identification and intervention as early as possible.

Back to the Table of Contents

Historical Highlights

The importance of identifying congenital hearing loss during the first few months of life was recognized more than 65 years ago. Ewing & Ewing say in their publication (1944): *“There is an urgent need to study further and more critically methods of testing hearing in young children. During the first year, the existence of deafness needs to be ascertained ... Training needs to be begun at the earliest age that the diagnosis of deafness can be established”*.

Audiometers were introduced in the 1940s, and they are a prerequisite for the professional fitting of hearing aids.

In 1965, the Babbidge report, issued by the US Department of Health, Education and Welfare, recommended the development and nationwide implementation of universally applied procedures for early identification and evaluation of hearing impairment.

The pioneering work of Marion Downs had a great impact on the process. Her findings have been broadly published since 1964.

In 1969, the Joint Committee on Infant Hearing (JCIH) was established with the goal of improving early identification of congenital hearing loss. At that time, however, appropriate hearing screening technology was not available. Therefore, the JCIH focused on screening only high-risk babies until their Position Statement in 1994.

In 1978, David Kemp (candidate for the Nobel Prize Award) in the UK measured otoacoustic emissions (OAE), which became one of the methods used for routine neonatal hearing screening. It is worth mentioning that this was not originally intended by him.

Interestingly, 1978 was also the year when the first multi-channel Cochlear implantation was performed in Melbourne, Australia.

Eight years later, in 1986 the first AABR (Automated Auditory Brainstem Response Equipment) became available.

During the years 1988 to 1999 there were several Federal Initiatives to support Early Hearing Detection and Intervention in the US. Position statements were published by the Joint Committee of Infant Hearing in 1994 and by the WHO in 1995.

The first European Consensus Conference was held in Milan in 1998 and resulted in a widely recognized Consensus Statement. This comprises 10 points and reads as follows:

- Permanent childhood hearing impairment (1) (PCHI) is a serious public health problem affecting at least one baby in one thousand. Intervention is considered to be most successful if commenced in the first few months of life. Therefore, identification by screening at or shortly after birth has the potential to improve quality of life and opportunities for those affected.
- Effective programmes of intervention are well established.
- Methods for identification of PCHI in the neonatal stage are now accepted clinical practice. They are effective and can be expected to identify at least 80% of cases of PCHI whilst incorrectly failing 2-3% of normally hearing babies in well-controlled programmes.
- Neonatal testing in maternity hospitals is more effective and less expensive than behavioural screening conventionally carried out at 7-9 months.
- Targeting neonatal testing on only the 6-8% of babies at increased risk (2) of PCHI reduces costs but cannot identify more than 40-50% of cases. Targeted neonatal hearing screening in parallel with 7-9 month behavioural testing is more expensive and less effective than universal neonatal screening.
- Hearing screening in the neonatal period cannot identify acquired or progressive hearing loss occurring subsequently. Surveillance methods are required to identify those cases, which may be 10-20% of all permanent childhood hearing impairment.
- Risks associated with neonatal hearing screening include anxiety from false positive results and possible delayed diagnosis from false negative results, but these risks are acceptable in view of the expected benefits.
- Neonatal hearing screening should be considered to be the first part of a programme of habilitation of hearing impaired children, including facilities for diagnosis and assessment.
- A system of quality control is an essential component of a neonatal hearing screening programme. Quality control includes training of personnel and audit of performance. The person responsible for quality control should be identified.
- Although the healthcare systems in Europe differ from country to country in terms of organisation and funding, implementation of neonatal hearing screening programmes should not be delayed. This will give new European citizens greater opportunities and better quality of life into the next millennium.

(1) Defined here as a bilateral permanent hearing impairment greater than or equal to 40 dB averaged over the frequencies 0.5, 1,2 and 4 kHz

(2) Examples include neonatal intensive care and family history of hearing impairment.

This NHS conference has been held biannually since then. The most recent one took place this year (in 2010).

The number of participants and countries they represent has increased continuously, and the successful implementation of screening programmes all over Europe has increasingly been brought to light.

[Back to the Table of Contents](#)



Dr. Dr. h.c. Monika Lehnhardt

Overview of the Worldwide Situation

The situation is still heterogeneous with many countries reporting 100% coverage, some with significant regional differences and some just at the verge of introducing NHS.

In **Canada** the situation is similar to some countries in Europe with regional differences. In the **US** we estimate that more than 90% of all newborns are screened.

Countries differ in the method they apply, whether it be OAE only, AABR only or BOTH OAE and AABR, which seems to have become “state of the art” and accepted practice.

In some countries, only one ear is tested, which seems insufficient in view of several studies documenting that children with single-sided deafness are facing additional difficulties at school and are delayed in their development.

In **Germany**, one of the most developed countries with a very high level of healthcare, it took nearly 10 years of lobbying until finally in early 2009, a law introducing UNHS was passed. The responsibility for implementation, however, lies with the federal lands, and varies greatly as a consequence.

As major obstacles to a high-quality implementation in general we identified lack of suitable equipment, insufficiently trained personnel, lack of central data processing and administration, insufficient counselling of parents and serious problems in financing.

Loss to follow-up varies dramatically from 2% to more than 50%, not only in the US but also in many European countries. A high percentage in loss to follow-up destroys the cost-effectiveness of the programme and can only be avoided through professional counselling of the parents.

[Back to the Table of Contents](#)

Definition and Quality of Screening

The definition for screening of the World Health Organisation reads as follows:

The main purpose of screening is the initial identification of undistinguishable diseases and disorders by using adequate methods. Screening allows selecting from the whole population patients with a high probability.

The WHO also defines the screening population as “*being special because society imposes care on asymptomatic persons who do not seek it*”.

What should be the criteria to decide in favour or against implementing any screening programme?

The American Academy of Paediatrics stated in the year 1999 that to justify universal screening, at least 5 criteria must be met:

1. An easy-to-use test that possesses a high degree of sensitivity and specificity to minimize referral for additional assessment is available;
2. The condition being screened for is otherwise not detectable by clinical parameters;
3. Interventions are available to correct the conditions detected by screening;
4. Early screening, detection and intervention result in improved outcome;
5. The screening programme is documented to be in an acceptable cost-effective range.

I believe the most important criteria are that screening should search for a disease or disorder which is frequent enough to justify the investment in time and money and – maybe even more importantly so – is treatable. Screening should be reliable, which means a very low incidence of false positive and false negative results.

In the case of screening for congenital hearing loss, all the criteria are fulfilled.

Deafness is the most frequent disorder in newborns.

Screening for congenital hearing loss is non-invasive, objective and non-expensive.

Adequate equipment to measure OAE and AABR is available.

Leading manufacturers state that the sensitivity of their equipment is 99% and the specificity 97%. This means we would have only 1% of false negative results (the baby is deaf but this was not confirmed) and 3% of false positive ones (the baby has normal hearing but we have a false alarm).

It is of paramount importance to keep these percentages as low as possible.



Dr. Dr. h.c. Monika Lehnhardt

The consequence of the false negative is that parents believe they have a normally hearing child, do not seek any treatment and communicate with their child in a way that is not adequate. It may well take up to 3 years for the hearing loss to become so obvious that parents insist on another assessment.

The consequence of a false positive result is that parents are shocked and will be under this impression until another assessment in a professional diagnostic centre corrects the result of the screening. This may take up to 6 months depending on local circumstances.

UNHS should be structured in such a way that all neonatal departments of hospitals are connected via Internet and deliver the data of their measurements to a central institution that collects all reliable epidemiological data and monitors the “tightness” of measurements (making sure that really all newborns are subject to screening) and the quality.

Key factors for a successful screening programme are suitable software to ensure quick and reliable data transfer, and adequate training for the personnel that implements the screening test (normally nurses in the neonatal ward).



Screening for congenital hearing loss should preferably be implemented on the 2nd or 3rd day after birth when the mother and baby are still in hospital. We know exceptions in countries like the Netherlands, where the majority of babies are born at home.

In case the baby fails to pass the test, a retest can be done on the following day.

Should the baby fail to pass again, she/he is referred to a Diagnostic Centre, where audiologists and/or ENTs provide

an in-depth analysis and identify the kind and level of potential hearing impairment.

As we all believe that family support is one of the most important factors in helping the hearing-impaired child to learn to listen and talk, we have to ensure professional and empathic counselling for the parents. We need to inform the parents about the results of the screening test and to explain what “pass”, “fail” and “referral” mean. We

Dr. Dr. h.c. Monika Lehnhardt

need to clarify the importance of the results versus alarm and the difference between screening and diagnosis.

In case the baby passes the test, we should not fail to indicate the possibility of a late onset of hearing loss and recommend that they insist on a new hearing screening at pre-school age. We should provide unbiased information about early intervention options and guide them to credible information about various approaches in rehabilitation.

Finally – and most importantly – we need to encourage family-to-family support by coaching parents on how to use modern ways of web-based communication (via Internet, Skype and LiveOnline rooms) to share knowledge and exchange experiences. With these modern tools of communication, families living in remote places can be in contact with other families in a similar situation, which is efficient and cost-effective.

[Back to the Table of Contents](#)



Dr. Dr. h.c. Monika Lehnhardt

NHS is the First Step Only

Once hearing loss is recognized, rehabilitation (more precisely, habilitation) should start immediately, i.e. even before the baby receives bilateral hearing aids, which should happen not later than 6 months after birth. Unfortunately, there are still many professionals who have no idea about the age when a hearing aid can be fitted. Many would fit the child far too late.



As we heard in the Conference of the European Hearing Aid Acousticians in Hanover this autumn, UNHS presents new challenges for the development of hearing systems and adaptation software for children who may now receive medical care between 0 and 6 months. Very small, yet child-safe dimensions as well as new acoustic couplings for the hearing system are required. Ear moulds that grow along with the child over a certain period of time and which reduce the risk of feedback are useful. Broadband frequency transmissions of up to 10 kHz is required for speech and language development.

Leading hearing aid companies like Widex (Denmark), Phonak (Switzerland) and Siemens (Germany) are actively working on this and presenting suitable hearing solutions for very small babies.

After a trial of 6 months with bilateral hearing aids, it should be clear whether the baby has any benefit from them or not. If this is not the case, parents should be offered the solution of a cochlear implant for their child. In the vast majority of European countries, in the US, Canada, Australia, this is considered a routine treatment and is funded by the Ministry of Healthcare or by insurance companies. More and more children receive bilateral cochlear implants based on scientific research that shows that children have additional benefit from the second implant. In some countries, bilateral cochlear implants have also become routine and are funded.

The quality, reliability and cosmetic appeal of cochlear implants have significantly improved since the first multi-channel implant in a 1 ½ year old girl was performed by Ernst Lehnhardt in 1988.

At that time, the average age of deaf born children to receive a CI was at around 5 – 6 years, but it has come down continuously ever since. Currently we aim at

implantation at the age of around 1 year. All leading companies in this field (Cochlear in Australia, Med-El in Austria and Advanced Bionics in US, and recently Sonova in Switzerland) offer high-quality products.

The reason for the dominant market share of Cochlear lies in their superior technology (they launched the new System 5 a year ago) and in their excellent service network.

Not only technology has changed to fit young children. Surgical techniques have also become less damaging to preserve any residual hearing. This is why indication has broadened, and children with residual hearing are also considered to be candidates for a CI.

With constantly improving results from the CI, some leading professionals feel confident in recommending a CI for single-sided deaf patients, when they do not benefit from cross-hearing aids or bone conduction hearing solutions. This is, however, still limited to adults.



We will see whether the vision of Dr. Jay Rubenstein expressed in 2004 will become reality: *“In 20 years, an implant will be the treatment of choice for anyone with a hearing loss greater than 50 dB”*.

[Back to the Table of Contents](#)

Conclusion

Universal screening for congenital hearing loss has been demonstrated to be feasible and cost-effective. Its success is dependent on sufficient educational effort on behalf of community professionals, commitment on the part of the programme planners, and data systems that track and recall infants who fail initial hospital-based screening.

One of the most decisive factors is counselling of parents in order to avoid loss to follow-up.

We also strongly advocate pre-school screening for hearing loss.

This is, unfortunately, not routine in the vast majority of European countries yet and it may take another couple of years of lobbying for pre-school hearing screening to become obligatory. Again, technology has advanced significantly and suitable equipment, such as the touch-screen *Senti* and *Sentiero* from Path Medical is available at a relatively moderate cost.

I conclude: Universal screening for hearing loss must be obligatory for newborns and I fully support what Martin Hyde says: *“Infants cannot complain of lack of hearing and cannot seek intervention we would surely not deny ourselves. We can and we must do their complaining for them. Infants and families have a fundamental right to early and effective communication”*.

[Back to the Table of Contents](#)

