Early detection of hearing loss

Abstract

The universal newborn hearing screening (UNHS) is currently spreading in Germany, as well, even though there can be no talk of a comprehensive establishment. The introduction of UNHS in several federal states such as Hamburg, Hessen, and Schleswig-Holstein can be ascribed to the personal commitment of individual pediatric audiologists. Apart from the procurement of the screening equipment and the training of the staff responsible for the examination of the newborns, the tracking, i.e. the follow-up on children with conspicuous test results, is of utmost importance. This involves significant administration effort and work and is subject to data protection laws that can differ substantially between the various federal states. Among audiologists, there is consensus that within the first three months of a child’s life, a hearing loss must be diagnosed and that between the age of 3 and 6 months, the supply of a hearing aid must have been initiated. For this purpose, screening steps 1 (usually a TEOAE measurement) and 2 (AABR testing) need to be conducted in the maternity hospital. The follow-up of step 1 then comprises the repetition of the TEOAE- and AABR measurement for conspicuous children by a specialized physician. The follow-up of step 2 comprises the confirmatory diagnostics in a pediatric audiological center. This always implies BERA diagnostics during spontaneous sleep or under sedation. The subsequent early supply of a hearing aid should generally be conducted by a (pediatric) acoustician specialized on children.

Keywords: Universal Neonatal Screening Program (UNHS), deafness, hearing disorders, tracking, screening, follow-up

1 Introduction

Congenital disorders represent the most frequent sensory malformation. Approximately one to two out of 1,000 healthy born babies (“well-babies”) will usually be affected; in case of children at risk (e.g. familial hardness of hearing, pre-, peri- or postpartal infections, premature birth with the baby weighing less than 1,500 g, transfusion-obliging hyperbilirubinamy, craniofacial dysmorphology etc.), a rate of one affected child per 50 newborns is to be expected [1], [2]. While the maturation of the visual pathway is completed during a child’s infancy at the age of approximately six months [3], the parallel process of the auditory pathway, similarly established at birth, extends over a period of four years. Since a constant adequate acoustical input is needed for the successful neuronal cross-linking that takes place in the course of the auditory pathway’s maturation, a well-functioning tympanum and inner ear are of utmost importance. In case of hardness of hearing, however, this crucial maturation of the auditory pathway cannot – or only insufficiently – proceed properly so that the development of the child’s hearing ability will be damaged irreparably. Without the ability to hear, a normal linguistic, intellectual and, consequently, social development is not possible [4], [5], [6], [7], [8]. Therefore, the late diagnosis of a hearing disorder stands in the way of a child’s normal intellectual and thus academic development in a way that from the age of four, even when supported optimally, the child will not be able to unfold its theoretical intellectual potential [8], [9]. That is why health professionals around the world try to examine the hearing ability of children only a few days after their birth so that, in case a child has been diagnosed with a hearing disorder, they are able to supply the hearing aid needed within the first three to six months of that child’s life [10], [11]. However, not every author finds support for the positive relationship between the early detection of an inner ear hearing disorder together with the resulting prescription of a hearing aid and the consequent positive impact on the linguistic development [12]. The IQWIG (“Institute für Qualität und Wirtschaftlichkeit im Gesundheitswesen” – Institute for Quality and Efficiency in the Health Care System) also proclaims that there is “indication but not proof that in a universal newborn hearing screening, identified children with hearing disorders have advantages in regard to their linguistic development” ([13], translated by the author). Nevertheless, a causal relationship is so evident for numerous experts worldwide that UNHS has already been established in many countries on several continents in a universal manner. In spite of a number of initiatives supporting a standardized screening [14], [15], [16], [17],...
area-wide UNHS is currently only offered in individual regions and federal states (see Figure 1) [23], [24], [25], [26], [27]. Many other countries – some of them in Africa [28], [29], the Middle East [30], [31] and the Far East [32], [33], [34], [35] – are momentarily trying to implement such a comprehensive universal newborn hearing screening (UNHS). Among them, there are developing and emerging nations [32], [35], [36] such as Malaysia [35], Thailand [32], Mexico [37] and India [38]. In Europe, various endeavors are being undertaken to introduce universal newborn hearing screening. Publications dealing with the subject not only originate in northern [39], middle [40], [41] and southern [2], [42], [43], [44] Europe, but also in, e.g., Serbia [45], Croatia [46], Albania [47] and Georgia [48]. A comprehensive UNHS can, among others, already be found in the following countries and continents: Europe (in Belgium, the Netherlands, Portugal, Austria [49], [50], Great Britain and Poland); in Australia, North America (USA [51], [52] and Canada), in Central America (Cuba) and in Asia (for example in Malaysia [35]). Some countries – for instance the People’s Republic of China – are currently preparing the implementation of such a nation-wide screening program. China works on a program that aims at combining visual and auditory tests [53] (see Figure 1). Austria performs screening of only one ear due to financial reasons (time saving during measurement). Studies proving that a one-sided hearing loss that has not been detected and, thus, not been treated can affect the social and cognitive development of children contradict this practice. Children suffering from undiagnosed one-sided hearing disorders run a significantly higher risk of school problems than children with normal hearing abilities [54], [55].

Programs for the establishment of UNHS have been on the rise for some years [32], [56], [57], [58], [59], [60]. An article published in the New England Journal of Medicine in 2006 already called it “a silent revolution” (“Newborn hearing screening – a silent revolution”) [61]. More than 20 years ago, Matschke and Plath, both from Germany, made suggestions for a serial examination regarding the hearing tests with children [62]. In more than 50% of all cases, the cause of congenital hearing loss are genetic disorders that, in turn, are mostly linked to GJB2 or GJB6 mutations [51], [63], [64], [65], [66], [67], [68]. A distinction can be made between syndrome-based and non syndrome-based hearing disorders [69].

2 Evidence of children’s hearing loss

The reason that a lot of effort is going into establishing UNHS is the frequency of occurrence of congenital hearing disorders. As described above, they are considered to be the most frequent congenital sensory disorder and more common than all other disorders for which screening blood tests are already being performed in newborns taken together. One out of 500 to 1,000 newborn children suffers from a congenital sensorineural hearing loss of at least moderate severity [4], [70], [71]. The number of first diagnosis of inner ear hearing disorders in Germany, a country without nation-wide UNHS (but various initiatives targeted at individual federal states [23], [24], [25], [26], [27], [72], [73] emphasizes how late a hearing loss is normally diagnosed when no comprehensive hearing screening is in place [74]. This is also true for our country where the density of physicians ranks high in a worldwide comparison and even though in Germany, children are looked at by a doctor in the course of the U exams as a matter of routine. Data from a study conducted by Finck-Krämer et al. (1998) reveal that the average age of chil-
children with inner ear profound hearing loss at the moment of diagnosis is 19 months, that children with severe hearing loss are app. two years old, that children with moderate hearing loss are four years old and that children with mild hearing loss are on average six years old at the moment of their first diagnosis [74]. The first suspicion of a disorder may have been aroused some months earlier – however, the first supply with a hearing aid took place even later [10], [74]. Yet, there is prove that the moment of first diagnosis can be accelerated significantly by means of UNHS [54], [75]. For instance, in the Hessian model in 2006, children at the moment of their first hearing disability diagnosis were, on average, only 3.1 months old [54].

3 Technical options for diagnostics

For years now, there have been a number of diagnostic methods for aural examinations even for newborns [76], [77], [78]. They allow for hearing ability testing of sleeping (or calm) children with objective measures in just seconds or minutes by staff that can be adequately trained in just a couple of hours and need not have any additional skills or may even be lowly qualified (and accordingly receive low wages). TEOAE- and AABR-examinations are used for this purpose for UNHS. DPOAE testing is not recommended in this context since the identified hearing threshold of this method is 40–45 dB, which may result in mild and moderate hearing loss remaining undetected [79]. (A description of the technical details of these methods will be omitted here. Corresponding literature can be found in the references.) A successful TEOAE test confirms a hearing ability which encompasses 30 dB, a successful AABR measurement certifies ability above 35 dB [77], [79].

Auditory neuropathies can only be accounted for by an AABR screening [80], [81]. With a percentage of 8.44%, that is in children with profound hearing loss, this disorder is much more frequent than previously assumed, as a study by Foerst et al. demonstrates [80]. Hence, for the aforementioned reasons, mild inner ear hearing loss cannot be ruled out even when the screening produces only inconspicuous results. That is why mild inner ear hearing loss is typically diagnosed later than more severe levels. Thus, the average age at the moment of the first diagnosis continues to be six years in Germany [74].

In their work published in NEJM, Paradise et al. came to the conclusion that untreated mucotympanones, another possible cause for mild hearing loss, do not harm the linguistic development substantially [82], [83]. This can also account for the late diagnosis of a mere mild hearing loss. Still, it must be pointed out that a reduction of the hearing ability by 30 dB is similar to using noise-suppressing ear plugs. A hearing threshold of 30 dB is equivalent to an increase in distance between somebody talking at normal volume and the target person by 30 meters [84]!

4 Follow-up costs of children's hearing loss

A consensus paper of the DGPP approved at the organization’s yearly conference in 2007 states that even a moderate threshold of 20–25 dB can indicate the necessity to try to adjust the hearing aid [85]. Children with moderate or higher degree hearing loss, however, will not develop normally in regard to their linguistic and intellectual abilities [86]. Consequently, during the first years of its life already, the time of the detection and treatment of a hearing loss determines the potential path a child with a hearing loss of higher degree may take in a society due to its educational background and professional qualification! This means that the early diagnosis of a hearing loss also has an impact on the economy as a whole: On the one hand due to the enhanced opportunity of development per se – on the other hand due to the costs that accumulate for medical treatment and differentiated types of school (regular or special school) [54], [87], [88]. The costs for a child with severe hearing loss accumulate to € 13,438, in case of risk screening only to € 8,241 and without systematic screening to € 4,760 [54]. So, at first glance, early detection is merely expensive. And in case a law was passed ordering the introduction of a nation-wide hearing screening, it is the health insurances that would have to raise the necessary funds. Yet, considering the macroeconomic costs for the welfare state by taking into account the educational costs for children in regular schools and for those in special schools dedicated to the therapy of speech and language difficulties, the contrary picture emerges. The education of a child with a hearing disorder during its first 16 years of life amounts to costs of € 125,778 in a UNHS program, € 140,605 for risk screening and € 155,944 without systematic screening. Accordingly, UNHS offers a saving potential of six to 15% concerning the education costs for children with hearing losses [54]. Even though in the short term, there will be an increase in costs, in the long run, savings can be expected [60], [87], [88]. In Germany, the short term increase in costs, however, would go to the detriment of the health insurance companies, as explained above. It is obvious that these companies will not benefit directly from the savings in the educational system. This is certainly one of the main reasons why in Germany, a nation-wide hearing screening for newborns has not been implemented, yet. Potential sources for the funding of such a program have not been identified [1], [89]. While there had been a hearing screening model project in the German federal state of Lower Saxony that had received its financing partly by health insurance provider AOK, this project does currently not receive any funding by public health care providers any more [90]. This is the case even though various expert commissions have been working on the launch of such a project for years and even though there is consensus about the usefulness and the necessity of such a program [4], [70], [71].
5 Personal commitment of many audiologists

It is thanks to the personal commitment of a number of physicians that in some federal states, a UNHS of assured quality could be implemented. As representatives for the many professionals who have rendered outstanding services to this matter, Markus Hess and Thomas Wiesner from Hamburg shall be mentioned here. In 2003, they were awarded the renowned “Hufelandpreis” for the first establishment of UNHS in a federal state, the city state of Hamburg. For the organization of a comprehensive quality-assured UNHS in Germany’s northernmost federal state Schleswig-Holstein [91], [92], Rainer Schönweiler from Lübeck even received two awards: the “Annelie-Frohn-Preis” of the German Association for Phoniatrics and Pediatric Audiology (“Deutsche Gesellschaft für Phoniatrie und Pädaudiologie” DGPP) in 2006 and the Federal Cross of Merit in 2007. Part of the reason why these awards were granted is that these pediatric audiologists, by means of immense personal commitment, succeeded in building the screening programs without being supported financially by health care providers. The financing for the technical equipment as well as for the personnel was raised mostly at trusts and private funds. The same is true for the efforts taken to establish a screening program in North Rhine-Westphalia, Germany’s most populous federal state; here, the pediatric audiologist clinics and departments of the University Hospitals of Aachen, Bonn, Cologne and Düsseldorf (ABCD-initiative) in the region North Rhine and the University Hospital of Munster as leader for the region Westphalia-Lippe are working intensively to implement a comprehensive UNHS. Also as representatives for many others who have proven outstanding dedication to the idea in the various federal states, Nawka in Mecklenburg-Western Pomerania, Gross in Berlin [70], [71], Strutz in Upper Palatinate [41], Neumann and Böttcher in Hesse [54] as well as Delb in Saarland [24], [73], [93], [94] should be mentioned. On a national level, Agnes Hildmann [71] is certainly the most influential advocate. What is distinctive about a comprehensive hearing screening, is not only that all maternity hospitals must be furnished with adequate screening equipment and that the staff who is in charge must be trained, but also that tracking – i.e. the follow-up on children with symptoms discovered during screening – must be ensured and that it can be guaranteed that these children will be examined more closely in specialized institutions in a timely manner in order to supply them with their first hearing aid until they are six months old [40], [70], [71]. For legal reasons, the parents’ consent is mandatory. In Hesse, in 0.6% of all cases, screeners were denied the permission of the parents – usually because the parents were subjectively sure that their children’s hearing was normal [54]. It is also important to confer with the data protection officials of the respective federal state. This is of particular importance with regards to the passing on of data to other federal states in the context of trans-regional screening programs (e.g. North Rhine-Westphalia and Hesse) [55].

6 Procedure for the implementation of a UNHS

When we are talking about the introduction of a comprehensive, quality-assured, universal newborn hearing screening today, we mean the following: A minimum of 95% of all newborns must undergo a hearing test; no more than 4% of all children tested ought to be conspicuous; it is necessary to register all children with moderate and severe hearing loss of this collective; the tracking of patients must be ensured, the prompt follow-up in specialized pediatric audiologist centers must be possible and an external quality control must be carried out [4], [70], [71], [95]. According to Schönweiler [92], the following steps are currently necessary when implementing a UNHS:

- In the respective federal state, a survey needs to be conducted to assess which clinics already offer hearing tests for newborns and which do not. It has to be asserted whether a sponsoring already exists.
- A political mandate needs to be developed while the further planning of the UNHS is initiated.
- A multidisciplinary consensus needs to be reached. This means that all groups of different professions involved (e.g. obstetricians, pediatrics, otorhinolaryngologists, pediatric audiologists, representatives of the nursing staff, commercial administration etc.) need to be brought together from the outset onwards.
- The process of the UNHS needs to be drafted. This process must include the registration with IT and data protection. Additionally, a research project together with an ethics proposal needs to be devised and it must be ensured that the screening devices as well as the education and training of the screeners result in an error-free feedback of the outcomes to the screening hub. It is also necessary that potential errors of the screeners themselves can be retraced at all times.

The phase of the actual execution of the UNHS necessitates the following activities:

- Information events and trainings for all actively involved screeners,
- Formation of a nonprofit sponsoring society for the purpose of raising funds; search for sponsors,
- Initiating public relations (WWW, press, radio, TV),
- Procurement of screening equipment for all maternity and pediatric clinics,
- Ensuring of a comprehensive post-screening and follow-up by training and certification of selected institutions (practices, clinics),
- Alignment of the registration and data protection with the specifications of the federal authority for data
protection, creation of a screening hub. It is only now that the registering can begin.

- Organization of recurrent trainings of all people involved, centrally as well as on site.

As a final requirement, the scientific analysis of results, publications among experts and the on-going optimization of the process are necessary [92]. The people responsible for the establishment of a UNHS need to decide whether they want to conduct a one- or two-step screening. A mere risk screening of children with the above mentioned risk factors, however, can result in 42% of children with profound hearing loss not being detected and is therefore considered not to be sufficient [96]. The two-step screening in Germany (e.g., the ABCD-initiative in the North Rhine region) for non-risk children (“well-babies”) includes TEOAE measuring first. Children with conspicuous results (pass criterion) need not be examined further while conspicuous children (refer criterion) undergo an AABR measuring [53], [54], [97], [98], [99]. Risk children, however, are directly tested using AABR and are also examined further using this method if the measure in the first trial resulted in “refer” (see Figure 2).

7 Risk factors for hearing disorders in children

The following characteristics are considered to be risk factors: familial hearing disorders, intratauterine infections caused by viruses during the first five months of pregnancy (for instance rubella, mumps or cytomegaly) and the treatment with ototoxic or teratogenic drugs. Similarly, oxygen deficits during birth, a birth weight below 1,500 g, an APGAR score below 4, a transfusion-obliging hyperbilirubinamy, a deformity of the head, craniofacial dysmorphies, syndromes/chromosomal aberrations, premature birth (<32nd week of pregnancy), intratauterine growth retardation, consanguinity of the parents, abuse by the mother, perinatal asphyxia, artificial respiration >10 days, severe respiratory adjustment disorder, intracranial injury or a postpartal meningitis [99], [100].

8 The “Hessian model”

The Hessian model, developed and established under the auspices of Katrin Neumann and Peter Böttcher, achieved a proven enhanced specificity from 95% to 97% by using the combined approach (two-step model) [54], [55]. The combination of both procedures also has additional advantages: The benefit of the TEOAE method is the shorter measuring period compared to the AABR method. Moreover, the detection threshold is 30 dB and thereby better than that of the AABR method by 5 dB. The costs for the consumable supplies are also lower [101], [102]. The AABR method, however offers better specificity and sensitivity [76], [103]. The two-step models are considered to be the most cost efficient [54], [104]. Vis-à-vis pure AABR-screenings, however, they have the disadvantage that auditory neuropathies may remain
undiscovered [80]. It is also for this reason that in North Rhine Westphalia, the region Westphalia-Lippe has decided to employ a pure AABR-screening. As for risk children, the likelihood of functional disorders above the outer hair cells – i.e. in the area of the acoustic nerve or the brain stem – is increased, their hearing ability is examined by using the AABR exclusively, also in the context of two-step screenings [54]. In rare cases, however, “well-babies” can theoretically, in spite of inconspicuous TEOAE-testing results, suffer from higher degree sensorineural hearing loss caused by an auditory neuropathy. The rate of auditory neuropathies in severe hearing losses is said to be higher even than 10% [80].

If the second step of a hearing screening examination, usually conducted by the maternity hospital, is conspicuous again, a further examination in a specialized institution should be facilitated within two weeks [4]. In the course of the follow-up steps 1 and 2 (see below), apart from binocularmicroscopic examinations, subjective testing based on free field audiometries, multi frequency tympanometric examinations, diagnostic OAE testing and BERA methods under sedation are applied [77]. Apart from Click-BERA-applications measuring the frequency range between 1,500 and 4,000 Hz, low chirp-BERAs (250–850 Hz) or notched noise-BERAs that are able to selectively investigate the frequency range of 500, 1,000, 2,000 and 4,000 Hz can be conducted. Infants that have been diagnosed with an inner ear hearing loss should receive a hearing aid between the age of three to six months prescribed by a specialized pediatric acoustician. The process of adaptation is dynamic. The pediatric acoustician and the pediatric audiologist accompany the affected children for years [11]. Half a year after the first hearing aid adaptation – i.e. ideally at the age of nine months – a BERA examination should be conducted since there are cases in which the hearing threshold apparently improves due to further ripening of the auditory pathway [105]. Thus, at the age of nine months, the BERA for control purposes serves as a means to virtually provide security for the diagnosis whereas the first BERA at the age of 3 months rather functions as a “working hypothesis”. If the hearing loss of the children is too significant to be sufficiently treated with a high performance hearing aid, a cochlea implantation (CI) even before their first birthday is indicated [106]. There are various specialized departments, mostly in university hospitals, for the subsequent rehabilitation. Hessian studies have shown that 56% of the children that had become deaf prelingually and had received a CI only after their third birthday could not attend a regular educational institution (regular kindergarten, regular school) whereas the percentage of children who had received the implant previous to their third birthday was only 24% [54].

In a study conducted in France, Koloski et al. investigated the point in time at which more children could be reached for a hearing screening; directly after birth at a hospital or two months later in the course of the U examination at the pediatrician. It appeared that 95.72% of the children in the maternity hospital, but only 64.18% of the children at the examination at the age of 2 months were tested [107]. Yet, further studies were also able to prove that hearing screening programs without quality-assured follow-up result in more than 50% of the children not going to continuing diagnostics, even when conspicuous in the first hearing screening test [108]. This is called the lost-to-follow-up-rate [54], [108]. It can be reduced to 7.8% (Hessian model) by means of a central registration of the children and guidance of the parents to adequate follow-up examination centers [54]. To achieve such low rates requires intensive activities in the screening hubs. Accordingly, Böttcher reports that in Hessa in 2006, 34,133 children were registered at the UNHS. Out of these, 2,220 needed to be tracked. For this purpose, the screening hub conducted 4,340 phone calls (out of which 956 concerned research at the maternity hospitals) and sent out 2,890 letters. The hearing loss of 66 children was confirmed [55]. This points out that a successful tracking with pleasantly low lost-to-follow-up-rates (7.8%) involves high administrative efforts and is very time consuming.

9 Projected UNHS process in North Rhine Westphalia

On the basis of the process of the hearing screening for newborns in North Rhine Westphalia (NRW) and of the position paper of the DGPP regarding the “Fundamentals of the quality assurance of a universal newborn hearing screening” published in 2007 [109], [110], the approach to the process of a hearing screening for newborns in a maternity and follow-up institution shall be depicted in the following (see Figure 1):

In the maternity hospital, the obligatory briefing of the expectant parents – orally as well as in writing – takes place. This is important since the support of the parents – especially in the case of conspicuous screening results – is crucial and increases the chance for a successful follow-up. Studies show that hearing screenings may well arouse fears on the part of the parents and that good information helps them to enhance their compliance and consequently reduce the lost-to-follow-up-rate [111], [112], [113]. Overall, the hearing screening of newborns is well supported by the parents [12]. Additionally, in Hesse and NRW, each child is assigned a multifunctional screening ID. This ID needs to be registered centrally once (for the ABCD-initiative, this is done through the Screening Hub North Rhine). The advantage of a screening ID compared to individual data entries is that it enables the automatic capture of personal data and avoids erroneous or multiple data entries. Moreover, screening IDs can for example be co-used for other forms of screening, e.g. for metabolic screening. After the parents give their consent to the screening and the testing methods, the results are documented in a yellow examination booklet and then, together with the screening ID, encrypted and transferred to the screening hub.
As to mature and healthy born infants, the hearing screening should take place prior to the discharge from the maternity hospital – shortly before prematurely born babies are discharged – yet, if possible, prior to the expected date of delivery [70].

If the screening result is conspicuous, the second step of the screening is to be undergone. The repeated hearing testing should preferably be conducted within four weeks by a resident phoniatrician/pediatric audiologist, or, respectively, by an ENT specialist or pediatrician who has a qualification in pediatric audiology (second screening). Children born by ambulant delivery, in birth houses or by home birth should also have their first hearing screening here. The same applies to cases where at the first testing, only one ear could be tested.

The quality of the screening devices is guaranteed by the manufacturer, whereas the operator is responsible for the regular maintenance and calibration of the equipment [110]. A pediatric audiological confirmatory diagnostic investigation (follow-up steps 1 and 2) is necessary in case the second step of the screening produced conspicuous results again.

If a child’s test results are conspicuous, the follow-up examination takes place during the stay in the hospital (second screening). If the result is continuously conspicuous, the first step of the two-step follow-up begins. In step 1 of the follow-up, the screening is firstly repeated in a special screening consultation with TEOAE- and AABR methods. Additionally, the findings for the ear are being collected binocularmicroscopically. If need be, additional examination methods such as tympanometry and free field measurement at the “Mainzer Kindertisch” will be applied.

In step 2 of the follow-up, a further pediatric audiological diagnostic investigation is carried out in a specialized pediatric audiological center. Here, confirmatory diagnostics, executed by BERA examinations, help to conclusively determine whether a hearing disorder is present that requires the supply of a hearing aid or not (see above). These diagnostics should be concluded within the first three months of a child’s life, the subsequent adaptation of the hearing aid within the first six months of a child’s life [70], [71], [110].

The screening that already takes place in the maternity hospital is performed by employees who have previously been taught successfully in a special training session of several hours. These training sessions are run by staff from the pediatric audiological centers (second step of the follow-up) or the screening hub. It is only the specially trained employees of the maternity hospitals who are authorized to perform the screening examinations of steps 1 and 2.

The screening for newborns in NRW maintains a transregional cooperation with the screening hub in Hesse. The data storage of the conspicuous testing or unscreened newborns (tracking by name) is located in Giessen. This central tracking office receives the tested children’s data of the associated screening institutions via an encrypted phone line on a daily basis (see Figure 3). To avoid measurement errors and incorrect entries, the data transfer of the personal data and data concerning the measurement or its quality is made directly from the measuring device (see Figure 4). Therefore, only those measurement devices are suitable that dispose of an input field as well as a modem for the transmission by telephone. Other measurement devices do not comply with the quality requirements of the screening for newborns in the ABCD-M initiative (Aachen, Bonn, Cologne, Dusseldorf and Munster). The data transmission of the follow-up institutions is managed via a browser-based VPN connection for which an internet server (follow-up server) was installed. It stores the incoming data in a central database. The data can only be accessed with the screening ID. Abuse of the data is thereby exacerbated to a maximum level. The tracking is supposed to prevent the loss of conspicuous children. Parents who do not comply with the screening will be contacted repeatedly by phone and also in written form and advised of the necessity of further diagnostics (see above) [55].

The previous education of the parents about the importance of the hearing screening for newborns aims at boosting their interest. Thereby, the lost-to-follow-up-rate can be drastically reduced. As mentioned above, this rate is below 8% in the Hessian newborn screening – American institutions, however, reach levels of considerably higher than 50% [54], [55].

The central tracking office administers additional controls for completeness and quality controls for screening and follow-up. For instance, it examines the testing quality of individual screeners, since the number of “refer”-trials as well as the number of test abruptions prior to successful “pass”-criterion for a child can be retrieved and checked. Moreover, the central responsibilities of the tracking office comprise the continuous mentoring and training of the screening personnel (see below), the counseling in case of technical difficulties, the coordination of the choice of measurement devices and methods as well as the monitoring of the process of confirmatory diagnostics in the follow-up institutions [109].

Apart from that, in the future, the screening offices will be responsible for the collection and archiving of data concerning the UNHS. It is planned that via the tracking office, participant and recall rates, rates of the children who have received follow-up, lost-to-follow-up-rates, rates of the children diagnosed with hearing loss, form, side and extent of the hearing loss, number of children in need of therapy and number of children who actually received therapy, date of diagnosis and therapy, and the rate of findings that have normalized over a period of several weeks and months will be collected and documented. Additionally, the regional screening headquarters shall be responsible for the collection of cumulative data such as the rate of hearing losses detected at a later point in time and the rate of the children born alive. The rate of conspicuous and inconspicuous findings, respectively, the overall coverage rate, the rate of false positive and false negative findings as well as the quality criterions of

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In step 2 of the follow-up, a further pediatric audiological diagnostic investigation is carried out in a specialized pediatric audiological center. Here, confirmatory diagnostics, executed by BERA examinations, help to conclusively determine whether a hearing disorder is present that requires the supply of a hearing aid or not (see above). These diagnostics should be concluded within the first three months of a child’s life, the subsequent adaptation of the hearing aid within the first six months of a child’s life [70], [71], [110].

The screening that already takes place in the maternity hospital is performed by employees who have previously been taught successfully in a special training session of several hours. These training sessions are run by staff from the pediatric audiological centers (second step of the follow-up) or the screening hub. It is only the specially trained employees of the maternity hospitals who are authorized to perform the screening examinations of steps 1 and 2.

The screening for newborns in NRW maintains a transregional cooperation with the screening hub in Hesse. The data storage of the conspicuous testing or unscreened newborns (tracking by name) is located in Giessen. This central tracking office receives the tested children’s data of the associated screening institutions via an encrypted phone line on a daily basis (see Figure 3). To avoid measurement errors and incorrect entries, the data transfer of the personal data and data concerning the measurement or its quality is made directly from the measuring device (see Figure 4). Therefore, only those measurement devices are suitable that dispose of an input field as well as a modem for the transmission by telephone. Other measurement devices do not comply with the quality requirements of the screening for newborns in the ABCD-M initiative (Aachen, Bonn, Cologne, Dusseldorf and Munster). The data transmission of the follow-up institutions is managed via a browser-based VPN connection for which an internet server (follow-up server) was installed. It stores the incoming data in a central database. The data can only be accessed with the screening ID. Abuse of the data is thereby exacerbated to a maximum level. The tracking is supposed to prevent the loss of conspicuous children. Parents who do not comply with the screening will be contacted repeatedly by phone and also in written form and advised of the necessity of further diagnostics (see above) [55].

The previous education of the parents about the importance of the hearing screening for newborns aims at boosting their interest. Thereby, the lost-to-follow-up-rate can be drastically reduced. As mentioned above, this rate is below 8% in the Hessian newborn screening – American institutions, however, reach levels of considerably higher than 50% [54], [55].

The central tracking office administers additional controls for completeness and quality controls for screening and follow-up. For instance, it examines the testing quality of individual screeners, since the number of “refer”-trials as well as the number of test abruptions prior to successful “pass”-criterion for a child can be retrieved and checked. Moreover, the central responsibilities of the tracking office comprise the continuous mentoring and training of the screening personnel (see below), the counseling in case of technical difficulties, the coordination of the choice of measurement devices and methods as well as the monitoring of the process of confirmatory diagnostics in the follow-up institutions [109].

Apart from that, in the future, the screening offices will be responsible for the collection and archiving of data concerning the UNHS. It is planned that via the tracking office, participant and recall rates, rates of the children who have received follow-up, lost-to-follow-up-rates, rates of the children diagnosed with hearing loss, form, side and extent of the hearing loss, number of children in need of therapy and number of children who actually received therapy, date of diagnosis and therapy, and the rate of findings that have normalized over a period of several weeks and months will be collected and documented. Additionally, the regional screening headquarters shall be responsible for the collection of cumulative data such as the rate of hearing losses detected at a later point in time and the rate of the children born alive. The rate of conspicuous and inconspicuous findings, respectively, the overall coverage rate, the rate of false positive and false negative findings as well as the quality criterions of
the screening can, in turn, be generated from this data. Cost-benefit analyses and the assessment of the efficiency of the UNHS could be retrieved from this data [110].

The funding of the described screening project in NRW (ABCD-M initiative) [109] is only secured for a period of two years. It is the objective to, in this time, with only two additional established posts and the honorary commitment of especially the senior employees of the pediatric audiological centers of the five university hospitals in NRW, establish a sound foundation for a comprehensive screening in Germany’s most populous federal state.

10 Closing words

Yet, only with political support will it be possible to establish a truly nation-wide and long-lasting UNHS in Germany. Hope remains that the personal commitment of many pediatric audiologists and ENT specialists has made it
possible that, by optimizing the tracking for UNHS, Germany will again be a pioneer in the successful hearing testing of newborns and infants.

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