


# BMJ Open Effectiveness and costs of a low-threshold hearing screening programme (*HörGeist*) for individuals with intellectual disabilities: protocol for a screening study

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## ABSTRACT

**Introduction** Individuals with intellectual disabilities (ID) often suffer from hearing loss, in most cases undiagnosed or inappropriately treated. The implementation of a programme of systematic hearing screening, diagnostics, therapy initiation or allocation and long-term monitoring within the living environments of individuals with ID (nurseries, schools, workshops, homes), therefore, seems beneficial.

**Methods and analysis** The study aims to assess the effectiveness and costs of a low-threshold screening programme for individuals with ID. Within this programme 1050 individuals with ID of all ages will undergo hearing screening and an immediate reference diagnosis in their living environment (outreach cohort). The recruitment of participants in the outreach group will take place within 158 institutions, for example, schools, kindergartens and places of living or work. If an individual fails the screening assessment, subsequent full audiometric diagnostics will follow and, if hearing loss is confirmed, initiation of therapy or referral to and monitoring of such therapy. A control cohort of 141 participants will receive an invitation from their health insurance provider via their family for the same procedure but within a clinic (clinical cohort). A second screening measurement will be performed with both cohorts 1 year later and the previous therapy outcome will be checked. It is hypothesised that this programme leads to a relevant reduction in the number of untreated or inadequately treated cases of hearing loss and strengthens the communication skills of the newly or better-treated individuals. Secondary outcomes include the age-dependent prevalence of hearing loss in individuals with ID, the costs associated with this programme, cost of illness before-and-after enrolment and modelling of the programme's cost-effectiveness compared with regular care.

**Ethics and dissemination** The study has been approved by the Institutional Ethics Review Board of the Medical

## STRENGTHS AND LIMITATIONS OF THIS STUDY

- ⇒ Multicentre screening study, comparing an outreach setting with a clinical setting.
- ⇒ Age-specific prevalence data of hearing disorders in a large sample of individuals with intellectual disabilities is collected in a population-based cohort study for the first time.
- ⇒ The effectiveness, as well as the cost and cost-effectiveness, will be analysed.
- ⇒ Follow-up period of 12 months allows a robust assessment of the clinical effectiveness of the programme.
- ⇒ The setting does not allow for a randomised control group and the participants are recruited from only one statutory health insurance (about 30% market share).

Association of Westphalia-Lippe and the University of Münster (No. 2020-843f-S). Participants or guardians will provide written informed consent. Findings will be disseminated through presentations, peer-reviewed journals and conferences.

**Trial registration number** DRKS00024804.

## INTRODUCTION

Individuals with intellectual disabilities (ID) are less healthy than the general society; in addition to syndrome-specific conditions, they are more likely to suffer from inactivity-related and lifestyle-related conditions, such as nutritional, dental, ophthalmic and cardiovascular diseases.<sup>1</sup> Many of these problems remain undiagnosed and untreated, not least because ID limits individuals' abilities



to communicate their health status and to participate in decisions about their own health and well-being. In addition, carers' knowledge of the medical history and potential health problems of care recipients is often inadequate. There is a need for people with ID to have external support in order to access health services.<sup>2,3</sup>

Hearing loss was ranked as the third most common cause of the loss of years of healthy life due to disability in the WHO's 2019 Burden of Disease Study.<sup>4</sup> The age-standardised prevalence was reported as 14.3% across all degrees of hearing loss and 3.5% for moderate-to-complete hearing loss in the European region.<sup>4</sup> For individuals with ID, the risk of being affected by persistent hearing loss is significantly higher than for the general population.<sup>1,5-7</sup>

Children with Down syndrome, for example, are frequently affected by recurrent otitis media, cerumen and related conductive hearing loss,<sup>8</sup> while persistent hearing loss is reported in 21%–50% of adults with the condition.<sup>9-13</sup> Age-related hearing loss (presbycusis) seems to appear in individuals with Down syndrome approximately three decades earlier than in the general population, while those with ID stemming from other causes experience presbycusis approximately one decade earlier than the general population.<sup>14</sup> Other syndromes related to ID, such as CHARGE, Wolf-Hirschhorn syndrome and Pierre Robin sequence, are associated with malformations of the ear canal or middle ear. Prenatal and postnatal infectious diseases, which are frequently associated with ID, greatly increase the risk of permanent hearing loss.<sup>15</sup>

Another challenge facing individuals with ID is that temporary or persistent hearing loss often remains undetected and undiagnosed in this population.<sup>1,7,16</sup> For example, 11.1% and 1.1% of people with ID who underwent hearing screening at two national Special Olympics Games (regional, national and international sports competitions for people with ID) in Germany were diagnosed with severe hearing loss or total deafness for the first time.<sup>17</sup> When hearing loss is diagnosed in this group it often remains undertreated or untreated.<sup>1,5-7,17,18</sup>

The scale and severity of hearing loss is clearly underestimated in individuals with ID. Its impact on this group is also significant. Undetected and untreated hearing loss can result in additional social and psychological struggles, social isolation, dementia and depression, and is associated with a reduction in quality of life.<sup>19-22</sup>

The prevalence and severity of hearing loss in people with ID is clearly underestimated and its impact on this population is significant. Unrecognised and untreated hearing loss in these individuals can lead to additional social and psychological problems, social isolation, dementia and depression, and is associated with a reduction in quality of life.<sup>22-25</sup>

In contrast, the early and continuous therapy of hearing disorders in people with ID is possible and beneficial.<sup>7</sup> Diagnosis and effective treatment of hearing loss at an earlier stage in individuals with ID can result in partial rehabilitation of hearing and communication

skills.<sup>23,24</sup> Shott *et al*, for example, reported that chronic middle ear infections in children with Down syndrome can be reduced by medication or surgical treatment to the extent that typical hearing levels can be achieved in 98% of cases.<sup>25</sup> Difficulties such as a lack of acceptance of hearing aids<sup>26</sup> and cochlear implants can be alleviated through early treatment and hearing rehabilitation.<sup>24</sup>

The treatment of older people with ID is also beneficial. For example, in a study by Evenhuis, the majority of study participants with ID over 70 years were able to use hearing aids without difficulty after individual habituation training.<sup>16</sup>

These findings clearly indicate that inadequately treated hearing loss is a relevant health issue in individuals with ID. One potentially valuable strategy for improving the diagnosis and treatment of hearing loss and its associated outcomes in this group could be regular hearing screening.

National and international consensus papers already recommend hearing screening in individuals with ID.<sup>27,28</sup> Further special recommendations have been developed for individuals with Down syndrome, who are disproportionately affected by hearing loss.<sup>29</sup> However, a survey by the European Federation of Audiology Societies Working Group on Audiology and Intellectual Disability indicated that these recommendations have not been implemented in most European countries.<sup>30</sup>

Simple, low-cost health screening for adults with ID has been repeatedly shown to have a positive impact on their health outcomes and on disease prevention, and was found to be less expensive than the usual care pathways.<sup>31</sup> Systematic hearing screening tests for individuals with ID were first conducted at the Special Olympics.<sup>1,32</sup> These screening assessments revealed that undiagnosed or inadequately treated hearing loss occurred in around a quarter of participants, and emphasised the need for regular hearing screening in those with ID. A follow-up of Special Olympics participants who had failed the screen and whose caregivers had received a recommendation that the care recipient see an otolaryngologist or hearing aid dispenser, however, revealed compliance in only 2% of cases.<sup>32</sup> One key reason for this, besides the frequent underestimation of the negative impact of untreated hearing loss by carers, seemed to be the effort required to organise such consultations. One solution to this problem might be to bring hearing screening programmes into the person's living environment (ie, their home, nursery, school or work).

### Aim of the study

The aim of the *HörGeist* study is to assess the effectiveness and costs of a low-threshold outreach programme for the identification and diagnosis of hearing loss within the living environments of people with ID, and their controlled assignment to therapy and therapy monitoring. The *HörGeist* programme is expected to result in a lower number of care recipients having inadequately treated hearing loss than spontaneous use of regular

care pathways. This is likely to lead to an improvement in hearing and communication skills and quality of life, factors which are to be assessed in this study. A cost-comparison between the *HörGeist* programme following failed screening tests and the regular care pathways will be conducted by economic modelling. Data on the age-dependent prevalence of hearing loss in individuals with ID will be collected. Coverage and compliance rates will be compared between outreach and clinical cohorts.

## METHODS AND ANALYSIS

### Study design

*HörGeist* is a screening study comparing the outcome of an outreach screening programme with clinical screening by postal invitation. The outreach cohort undergoes hearing screening in the participants' living environments ('come strategy') and, if hearing loss is suspected, further diagnostics or review of any already-ongoing therapy and, where necessary, the initiation of on-site therapy followed by structured monitoring. If on-site diagnosis or treatment is not possible, participants are referred to local practices or hospitals. Members of the control group receive an invitation letter via their families to participate in the same procedures but within a clinical setting ('go strategy').

The study is funded by the German Innovation Fund of Germany's Federal Joint Committee (01NVF18038).

### Study setting

This multicentre study is coordinated by the Department of Phoniatics and Paediatric Audiology, University Hospital Münster, Germany (principal investigator (PI): Katrin Neumann). It is conducted in cooperation with the Chair of the Department of Special Education and Rehabilitation, Division of Audio-Pedagogy, University of Cologne. The screening centres used for the clinical group are phoniatics and paediatric audiology departments or practices at the Universities of Cologne and Essen, and in the city of Düsseldorf. Investigators at all centres specialise in phoniatics and paediatric audiology and are experienced in the diagnosis and treatment of hearing loss in people with ID.

The recruitment of participants in the outreach group will take place within 158 institutions that form the living environments of individuals with ID. Depending on the age group, the hearing screening assessments will take place in nurseries, day-care centres, schools, living facilities and workplaces. The type and planned number of facilities and participants are outlined in [table 1](#).

### Participants and recruitment

A total of 1191 individuals who meet the inclusion criteria (infants, children, adolescents and adults with ID who are insured with the AOK Rheinland/Hamburg, the largest statutory health insurer in the region) will be recruited for the study. Only individuals for whom a hearing test poses a risk to themselves or the examiners are excluded. For the

**Table 1** Type and planned number of facilities and participants to be recruited

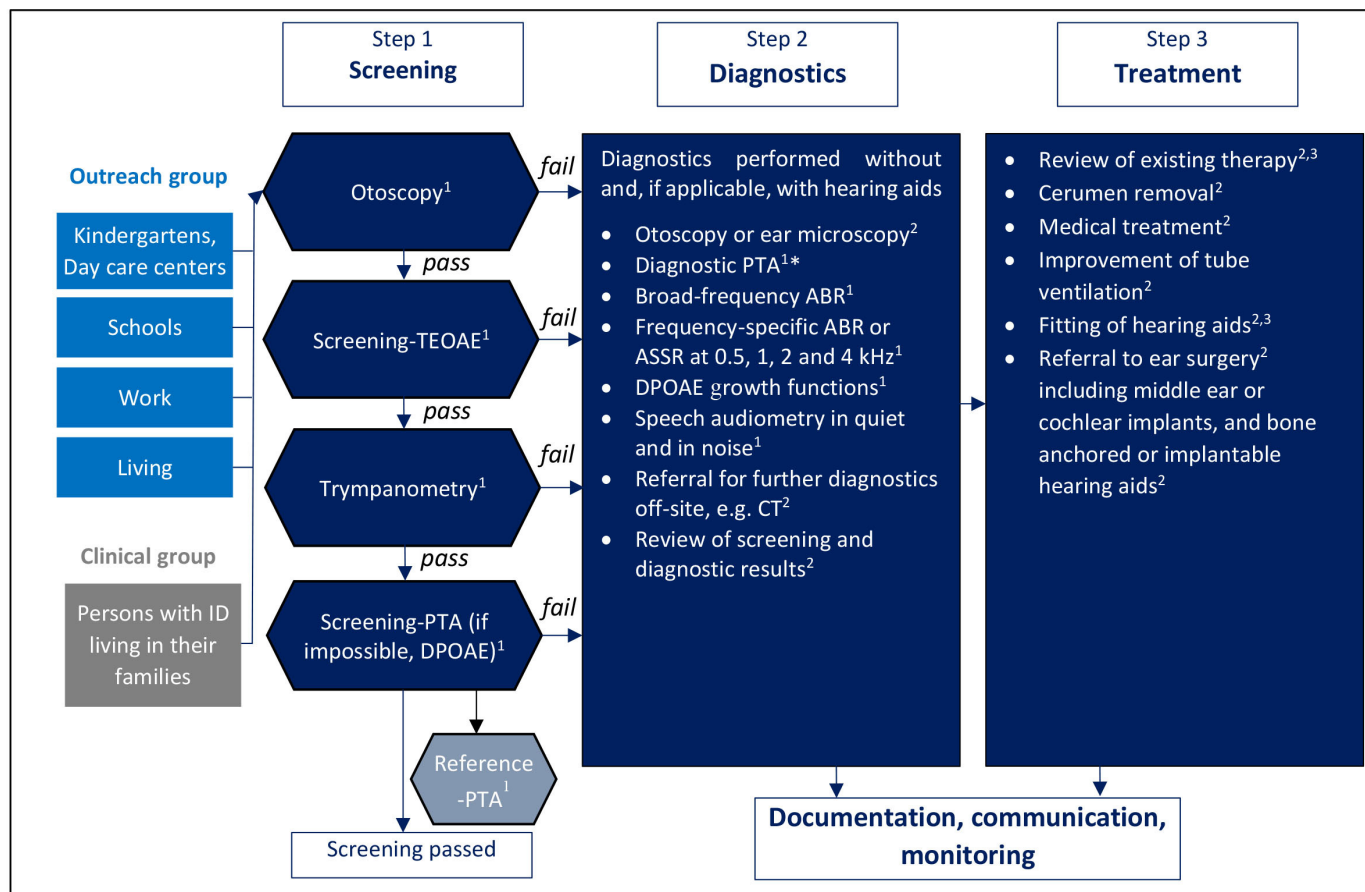
	Planned no of facilities	Planned no of participants
<b>Infants and children (age 0 to &lt;6)</b>	<b>46</b>	<b>350</b>
(A) Special education nurseries	20	190
(B) Day-care centres with integrative groups or regular nurseries/day-care centres with individual integration	20	70
(C) Residential facilities for children with ID	6	90
<b>Children and adolescents (age 6 to &lt;18)</b>	<b>29</b>	<b>350</b>
(A) Special schools	10	250
(B) Regular schools with inclusion of children or adolescents with ID	15	40
(C) Residential facilities for children and adolescents with ID	4	60
<b>Adults (age ≥18 years)</b>	<b>83</b>	<b>350</b>
(A) Sheltered workshops	3	100
(B) Company-integrated workplaces	50	50
(C) Residential facilities for adults with ID and assisted-living facilities	20	140
(D) Outpatient living facilities	10	60
Bold values show the sum of the subgroup values listed below. ID, intellectual disabilities.		

outreach group, 1050 participants will be recruited across three age groups (0 to <6 years, 6 to >18 years, >18 years) through the facilities listed in [table 1](#). For this, all facilities in the catchment area of the AOK Rheinland/Hamburg listed in [table 1](#) (the city of Hamburg itself is excluded because it is too far away from the Rheinland region) will be invited in writing and by telephone to participate in the *HörGeist* programme. The heads of facilities who are interested in participating will receive detailed telephone advice from the project management team and written study information. They then select potential participants who are insured with the AOK Rheinland/Hamburg, check their eligibility for inclusion and distribute information material and consent forms (online supplemental annex 1 and 2) to them and/or their parents or legal guardians (written in simple language for persons with ID ≥10 years old). If consent is obtained for a person with ID, the facility leader will inform the study team who will then then organise a screening appointment. The study physician will receive written informed consent from the participant and/or their parents or legal guardian prior to the appointment.

For the clinical control group, 141 AOK-insured individuals with ID will be randomly selected by the health insurer and invited by post via their families to hearing screening tests in one of the nearby phoniatic-paedaudiological departments or practices belonging to the study centres.

The first patient was included in the study on 30 September 2021. The follow-up of the last patient ends on





**Figure 1** Flow chart of the three-step *HörGeist* screening programme. <sup>1</sup>Screening staff; <sup>2</sup>Physician; <sup>3</sup>Hearing aid acoustician; \*In case of failed screening, reference PTA is the diagnostic PTA. ABR, auditory brainstem response; PTA, pure tone audiometry.

30 September 2023. The collection of cost data continues until 30 June 2024, due to the large delay in claims data of statutory health insurances in Germany.

### Screening programme

The *HörGeist* programme (figure 1) for both the outreach and clinical groups, consists of a hearing screening assessment and, if a hearing loss is indicated, immediate further diagnostics and initiation of treatment or optimisation of an already existing treatment. This is carefully documented in case report forms (CRFs) for each individual.

The first step of this programme, the hearing screening assessment (figure 1, step 1) includes (1) otoscopy, (2) transitory evoked otoacoustic emissions screening, (3) tympanometry and (4) pure tone audiometry (PTA) screening at 0.5, 1, 2, 4 and 8 kHz (air conduction) performed either in the classical manner or using an adaptive self-test (Multiple-choice Auditory Graphic Interactive Check). If PTA is not possible, distortion product otoacoustic emissions (DPOAE) are recorded at 1, 2, 4, 6 and 8 kHz. A reference PTA with additional threshold measurements for air and bone conduction will also be conducted in order to assess the validity of the screen. If participants fail one or more of the screening tests, immediate audiological diagnostics will follow at the same facility (figure 1, step 2).

A detailed medical history will be taken by two doctoral students and the screening staff prior to the screening assessment, using a self-developed questionnaire (the authors are not aware of any comparable tools) covering previously known and, where applicable, treated hearing loss, previous ear surgeries, hearing aids worn, other previous illnesses (especially those that are comorbidities of hearing disorders), surgeries, medication taken, lifestyle habits (such as smoking and alcohol consumption), hearing-related quality of life and communication skills. Blood pressure, height and weight are also measured in order to help identify risk factors for hearing impairment.

Further diagnostics include: (A) otoscopy or ear microscopy conducted by the study physician; (B) diagnostic PTA (identical to the reference PTA); (C) speech audiometry in quiet and noise using either the Freiburg monosyllabic test or the Mainzer Audiometric Test for CHildren (MATCH); (D) (if PTA is not possible or the results are unclear) auditory brainstem response (ABR) using broadband click or chirp stimuli, and frequency-specific ABR or auditory steady state response at 0.5, 1, 2 and 4 kHz and (E) (if PTA and ABR are both not possible) hearing threshold estimation on the basis of DPOAE growth functions.

The screening assessment and further diagnostics are supervised by the study physician for the outreach group. The

physician inspects the ear canals and eardrums using televideootoscopy where necessary, evaluates the audiograms, and advises the screening staff and the participant on the further procedure. They also prepare a doctor's letter for each participant's general practitioner and for the parents/guardians, in which the findings and recommendations are reported. If therapy is necessary, the study physician may offer to initiate it directly at the screening facility, for example, cerumen removal, prescriptions for drug treatment or nasal balloon treatment in cases of dysfunctional middle ear ventilation. Hearing aid fitting can also be initiated by the study physician in cooperation with a hearing aid acoustician within the study team (figure 1, step 3).

If further diagnostics cannot be completed locally, the study physician will recommend their completion at one of the project-affiliated phoniatic-paediatric study centres. Attendance at other centres (phoniatic-paediatric clinics, ear, nose and throat clinics/practices or hearing aid acousticians) may be recommended by the study physician, for example, where ear surgery is necessary, if existing therapy needs review and optimisation, or where the parents/carers wish to have therapy elsewhere. The progress of these externally performed diagnostics and treatment will be monitored by telephone interviews conducted by two doctoral students.

Diagnostics and treatment for the clinical group will be provided by one of the study centres, with referral to

other aforementioned facilities if necessary, and will also be monitored by letters and telephone.

The *HörGeist* screening programme will be repeated for each participant after 12 months in order to check the results of the previous year's screen, check therapy outcomes where necessary, detect newly developed hearing losses, and thus to be able to estimate the optimum time period for follow-up screening.

### Outcome assessment

Figure 2 is a sketch of the study design showing the outcomes and measurement time points before screening ( $t_0$ ) and at follow-up 12 months later ( $t_1$ ).

### Primary outcome

The primary outcome is the difference between the proportion of participants/ears with inadequately treated or untreated hearing loss out of the total number of participants/ears, between the first ( $t_0$ ) and the second hearing screening ( $t_1$ ).

### Secondary outcomes

Overall and age-associated, type-associated, side-associated, severity-associated and environment-associated prevalence of hearing loss.

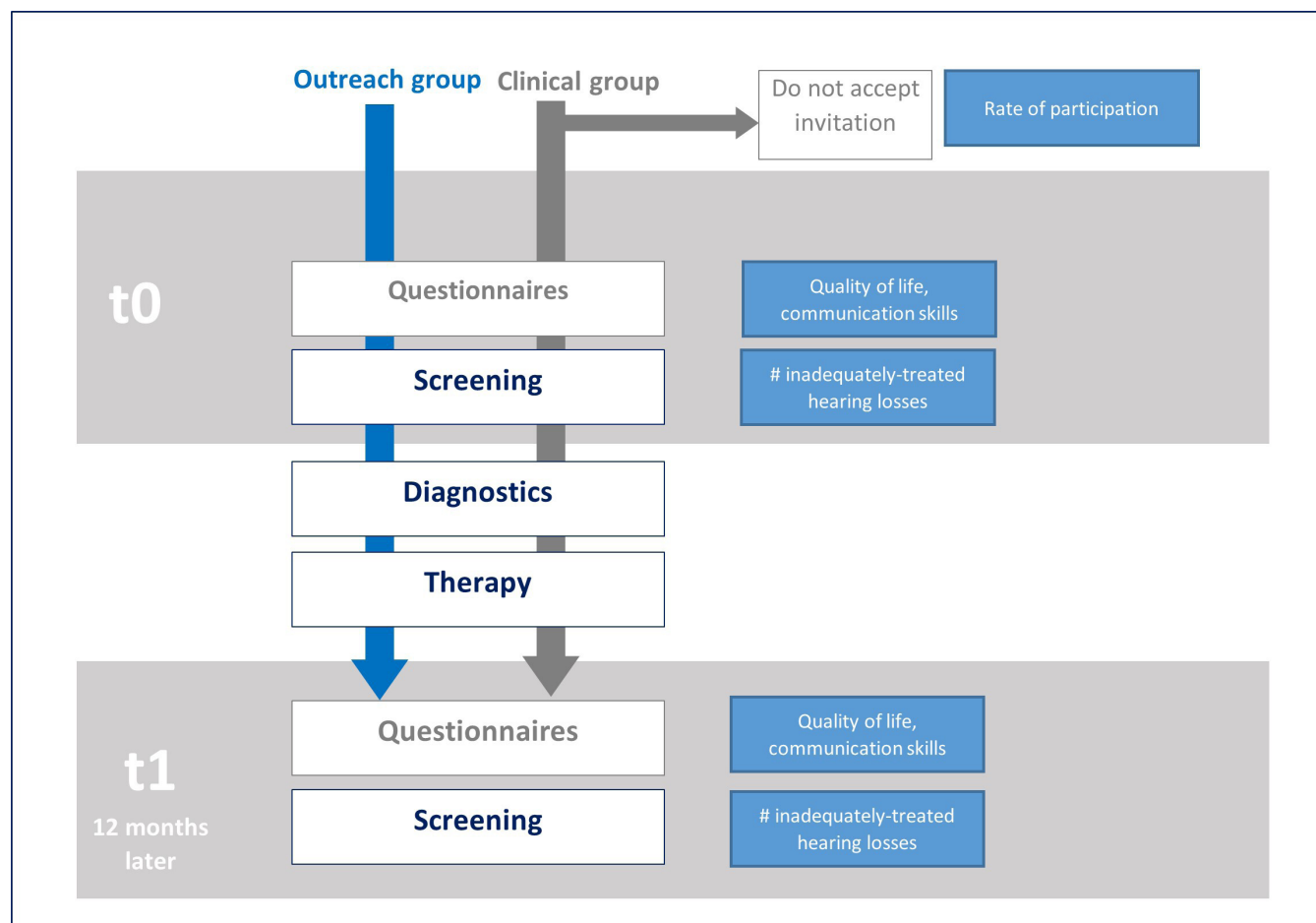


Figure 2 Study protocol and outcome assessment.

Hearing-related quality of life, communication skills and comorbidities of the participants as assessed by questionnaires administered to parents, relatives, carers or guardians

Coverage and compliance rate, specificity, sensitivity, negative and positive predictive values of the hearing screening assessments.

Cost data/resource utilisation of the *HörGeist* screening programme as assessed with a standardised questionnaire. Information will be collected regarding ongoing programme costs (eg, personnel costs for all programme steps that are not part of the regular care pathways, material and travel costs for screening) as well as one-time costs for programme design and implementation.

Cost of illness 12 months before-and-after enrolment in the programme will be calculated based on claims data of a large statutory health insurance company.

Cost-effectiveness of the programme compared with regular care will be conducted by health economic modelling.

### Sample size

The McNemar test will be used with a Bonferroni-adjusted significance level of 1.67% (for an overall level of 5%) to assess the difference in the number of inadequately treated hearing losses (primary endpoint) in each of the three age groups of the outreach group. The sample size calculation was performed for a power of 80%. The amount of inadequately treated hearing loss is expected to reduce from 7.5% to 3.5% over the period t0–t1. The initial 7.5% is a conservative estimate based on the expected prevalence of hearing loss (15%) and the high proportion of undiagnosed hearing loss in people with ID (11%).<sup>17</sup> With an assumed screening refusal rate at t0 of 3% and a drop-out rate of 5% (based on several thousand hearing screenings during Special Olympics games<sup>16 7 32</sup>), approximately n=350 individuals with ID per age group need to be included. The characteristics, as well as the share of inadequately treated hearing loss at baseline, will be compared between drop-outs and non-drop-outs. The sample size for the clinical control group has been calculated for the comparison of the utilisation rate between outreach and control groups. Assuming utilisation rates of 97% and 85% (outreach and control groups, respectively), Fisher's exact test provides a power of 80% if 47 individuals per age group (141 in total) are invited into the control group.

### Data analysis

#### Clinical evaluation

The primary outcome in the three age groups is the difference in the number of inadequately treated hearing losses between initial screening, diagnostics and treatment, and repeat measurements 1 year later. The difference will be described in each age group using a contingency table and tested by Bonferroni-adjusted McNemar tests with an overall significance level of 5%. The number of individuals who use the programme and the number of those

who adhere to it will be compared between the outreach and control groups.

#### Data merging

The study data will be forwarded to the Competence Centre for Clinical Studies Bremen (KKSB). This centre will transmit a list of AOK-insured participants (name, date of birth, insurance number) and corresponding pseudonyms to AOK Rheinland/Hamburg. Further health-relevant data will be requested from AOK and transferred in pseudonymised form to the trust centre of the KKSB. All health-relevant data, questionnaire data and data from hearing screening, diagnostics and therapy will be merged with the claims data and used for further analysis.

#### Health economic evaluation

The AOK Rheinland/Hamburg—a large statutory health insurance company with which approximately one third of the total population in the study region (Rheinland, Germany) is insured—will provide the claims data of its insured study participants. These data will include health-care utilisation and expenditure for each individual in the 12 months prior to enrolment in the study and in the 12 months from t0 to t1. The claims data and the clinical data will also be merged by the trust centre of the KKSB.

A calculation of the total costs will be carried out from a statutory health insurance perspective. Net costs without co-payments will, therefore, be considered. The data will contain information on inpatient hospitalisation costs, outpatient hospitalisation costs, outpatient healthcare costs, costs for pharmaceuticals, remedy costs and costs for rehabilitation (those paid by the health insurance, as some rehabilitation measures are paid by other social insurances). The total healthcare insurance costs will be calculated for the 12 months prior to and after enrolment to compare the cost of illness before-and-after programme participation. Subgroup analyses will be carried out where possible, for example according to age, gender, life environment, kind and severity of hearing loss, in order to determine whether cost of illness is different in the corresponding groups. Sensitivity analyses will also be performed. The evaluation will be carried out following the recommendations of the 'Good practice secondary data analysis'.<sup>33</sup>

A microcosting analysis of the programme cost will be carried out. A standardised questionnaire will be developed for this, which aims to capture the resource use associated with screening and diagnostics in the *HörGeist* programme.

#### Markov model

In a Markov model, patients undergo different, mutually exclusive health states in one or more time intervals each of 1 year's length (Markov cycles).<sup>34</sup> The Markov model will be developed to analyse the cost-effectiveness of the *HörGeist* programme versus regular care in detecting hearing loss in individuals with ID. Study data relating to

screening results, costs of illness in the outreach group and the programme cost will be implemented into the model. Systematic literature research will be conducted in order to feed the model with further relevant data for regular care regarding cost of illness and treatment. Sensitivity analyses will be done to account for uncertainty. Relevant parameters of the model will be varied and the impact on overall results assessed.

### Patient and public involvement

A large statutory health insurance was/is involved in the design, conducting, reporting and dissemination plans of this study. Extensive discussions and information of the patients, legal guardians and facilities were conducted before starting the programme.

## ETHICS AND DISSEMINATION

### Ethical issues

The clinical protocol and written informed consent were approved by the Institutional Ethics Review Board of the Medical Association of Westphalia-Lippe and the University of Münster (approval number 2020-843 f-S). All procedures described in the protocol follow the Good Clinical Practice guidelines and the ethical principles described in the current revision of the Declaration of Helsinki.<sup>33 35</sup> All local legal and regulatory requirements will be fulfilled.

The main ethical issues are informed consent, the use of the *HörGeist* programme, the protection of data privacy and the inclusion of underage as well as incapacitated participants with the consent of their parents or legal guardians.

All individuals approached will receive detailed information and explanation in simple language regarding the study protocol and the *HörGeist* programme before enrolment. Written consent must be given by each participant or their parent or legal guardian.

Contact addresses will be provided for further questions regarding participation in *HörGeist* or in cases of withdrawal.

### Dissemination plan

The main results will be published in a final report, as required by the German Innovation Funds directive. The scientific results will be disseminated via articles submitted to peer-reviewed scientific journals following the International Committee of Medical Journal Editors authorship eligibility guidelines, and via presentations at national and international scientific conferences. The *HörGeist* manual will be published in detail at the end of the project.

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