HEARING IMPAIRMENT IN ADULTS WITH AN INTELLECTUAL DISABILITY: EPIDEMIOLOGY AND REHABILITATION

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HEARING IMPAIRMENT IN ADULTS WITH AN INTELLECTUAL DISABILITY: EPIDEMIOLOGY AND REHABILITATION

Slechthorendheid bij mensen met een verstandelijke beperking: epidemiologie en behandeling

Proefschrift

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aan de Erasmus Universiteit Rotterdam
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mijn moeder
mijn vader †

CONTENTS

Chapter 1

INTRODUCTION

BACKGROUND

Hearing impairment is an invisible problem which may remain hidden, especially in persons who are not able to realise and tell that they are hearing poorly. This is the case in early childhood, but also in many persons with an intellectual disability (ID). For the detection of the hearing impairment they depend on their carers who may not recognise the hearing impairment. It is possible that in persons with an ID, signals of hearing loss are interpreted by carers as consequences of impaired cognitive abilities. In this way a lack of response, inadequate reactions to sounds, or odd reactions during a conversation can be interpreted erroneously.

Generally, undetected hearing impairment in early childhood can lead to delays in the development of speech and hearing, and herewith to cognitive impairment, as well as to social and emotional problems. It is therefore crucial that hearing impairment is detected and treated at an early age.[1,2,3] If the child also has an additional intellectual disability, the consequences of the hearing impairment will be aggravated due to a lack of compensatory abilities. Because of this, treatment has to be considered at a lower level of hearing impairment than for people with normal intellectual capacities.

When a more severe hearing impairment in adults with an ID has been present from early childhood and remained undetected, and so not treated, it will have hampered communication skills and intellectual progress. This in addition can negatively influence the level of functioning, and herewith increase the dependence on carers. It is even thinkable that persons who are now classified as intellectually disabled would not have been so, if hearing impairment would have been detected and treated at an earlier age.

Hearing loss in adults may result in feelings of anxiety, stress, and isolation.[4] Cognitive limitations may intensify these feelings because they increase the inability to comprehend and predict surrounding events. Because of this, behavioural problems may arise of which the cause - the hearing impairment - may not be recognised. It may be possible that treatment of the hearing impairment, reduces the behavioural problems.

In the Netherlands, population screening of hearing function was introduced around 1965.[5] It was performed in children at the age of nine months by means of behavioural screening with distraction methods.[6] Unfortunately children with developmental delays had to be excluded from this hearing screening because they are unlikely to show age-appropriate auditory maturation responses.[3] Many of these children were followed up by paediatricians who in the past quite often did not refer these children for assessment of the hearing function.

At the end of the last century this situation changed for the very young children by the introduction of neonatal hearing screening. No children are excluded from this screening programme.[5]

In adults with an ID, until recently not much attention has been paid to hearing impairment. This applies to screening and diagnosis, as well as to audiological rehabilitation. Guidelines for early identification and diagnosis of hearing impairment adapted to persons with an ID were needed. These were developed in

1995 by a working group consisting of representatives of the Netherlands Society of Physicians for People with Intellectual Disabilities (NVAVG), audiologists, ENT specialists, general practitioners, behavioural scientists, as well as of representatives of the Speech and Language section of the Dutch Society for Promotion of Expertise in the Care of Subjects with Intellectual Disabilities (NGBZ), and of the Dutch Institute for Health Care Improvement (CBO).[7,8] Hearing impairment was defined as a hearing loss of 25 dB and over at the best ear, averaged over 1, 2 and 4 kHz. Later, international consensus was developed on early identification of hearing and visual impairments in children and adults with an intellectual disability by the Special Interest Research Group of the International Association of Scientific Studies on Intellectual Disability (IASSID).[9]

The next step was to estimate the size of the problem of hearing impairment in adults with an ID in the Netherlands, its relation to risk groups, and the proportion of hearing impairments that had remained unnoticed. The first part of this thesis aims at answering epidemiological questions like: will the study results support the IASSID guidelines for early identification, are adjustments in these guidelines necessary?

The second part of this thesis focuses on audiological rehabilitation in adults with an ID. Our clinical experience is that this often fails. This may be caused by client related factors which have to do with different expectations of adults with an ID towards hearing aids as compared with the general population, or with different experiences with the devices once they have been introduced. Obviously just handing over the hearing aids with some instruction to the client and carer is not enough. We need to know what elements a treatment protocol should consist of to provide optimal audiological rehabilitation in adults with an ID. Subsequently we have to investigate the feasibility of the implementation of such a protocol in ID services, and to identify the factors that influence the implementation process.

In the next paragraphs of this introduction, after a section with definitions, previous study results on these topics will be presented, the Dutch situation of audiological care for the ID population described, and aims of the studies formulated.

DEFINITIONS

1. Degree of intellectual disability was classified as follows: mild: IQ 55 - 70, moderate: IQ 35 - 55, severe: IQ 25 - 35, profound: IQ below 25.
2. Hearing impairment was defined, according to the criteria of the World Health Organization[10] as loss of more than 25 dB at the best ear, however not averaged over four frequencies (0.5, 1. 2 and 4kHz) as in the WHO definition, but over three (1, 2 and 4 kHz) according to the International Consensus Statement of the International Association for the Scientific Study of Intellectual Disabilities.[9] The degree of hearing impairment was classified as: mild loss: 26 – 40 dB, moderate loss: 41 – 60 dB, severe loss: 61 – 80 dB, profound loss: over 80 dB (WHO 1997, classification for adults).
3. Visual impairment was classified according to the WHO-criteria:

visual impairment: visual acuity < 0.30, but not < 0.05, and/or visual fields < 30°, but not < 10°.
Blindness: visual acuity < 0.05 and/or visual fields < 10°.

4. Dual sensory impairment (DSI) was diagnosed in case of a combination of visual and hearing impairment, as defined above.
5. Implementation: the systematic introduction of innovations and/or changes of proven value, with the aim of structural embedding in professional behaviour, in the functioning of (an) organisation(s), or in the structure of health care.[11]

PREVIOUS EPIDEMIOLOGICAL STUDIES

Not many epidemiological studies of hearing impairment in the population with intellectual disabilities are performed. It has been shown that children with Down syndrome are at an increased risk of hearing impairment due to chronic or recurrent middle ear infections.[12,13] An increased risk of hearing impairment has also been shown in adolescents and (young) adults with Down syndrome.[14,15,16] In the studies in adults it appeared that, apart from the conductive losses due to chronic middle ear problems, progressive inner ear hearing loss with characteristics of age-related hearing loss was identified even starting before the age of 30 years.

On prevalence of hearing impairment in persons with an intellectual disability by other causes than Down syndrome very little is reported. Screening results were published of a small population in an English day-care centre[17] and a series of Dutch community-based homes,[18] as well as in a large Australian population[19] identifying hearing impairment in 25 - 42% of the people. Unfortunately, definitions of hearing impairment in these studies were varying, and prevalences, at least in the English and Dutch study, were valid only for selected community-based populations. In the USA,[20] and in Israel[21] hearing function in populations of 40 years and older was investigated amongst other health problems. This was done by means of questionnaires, showing respectively 27% and 20.4% of cases with hearing impairment. Schrojenstein Lantman-de Valk et al[22] performed a large Dutch population-based study of a range of physical conditions, including (undefined) hearing impairment, but this study was also based on questionnaires, not on assessments. Based on the aforementioned Dutch consensus guidelines,[8] the total population of a Dutch institute was assessed, identifying prevalences of hearing impairment increasing from 21% in the subgroup younger than 50 years with intellectual disabilities by other causes than Down syndrome up to 93% in those aged 50 years and over with Down syndrome.[23] Again, this was a selected population.

These studies show that people with intellectual disabilities are definitely at an increased risk, but none of them was designed well enough to provide valid population prevalences. Neither did they show, whether the risk was increased in all subgroups, including the largest one: those aged younger than 50 years with mild intellectual disabilities by other causes than Down syndrome. Because of this, in 1998 we started a cross-sectional epidemiological study on hearing impairment in a population, representative for the Dutch adult population of intellectual disability (ID) service users

Aims of the epidemiological study

The aims of this epidemiological study were to assess prevalence and severity of hearing impairment in Dutch adult ID-service users, and the relation of the prevalence of hearing impairment to age, Down syndrome and severity of ID. Another aim was to assess the percentage of cases in which hearing impairment had not been identified prior to the study (**chapter 2**).

Hearing function as well as visual function were assessed in the same study population. Outcomes of visual assessments have been reported elsewhere.[24,25,26,27] Now that hearing assessments were completed as well, an additional aim could be realised: assessment of the epidemiology of dual sensory impairment in this population (**chapter 3**).

PREVIOUS STUDIES ON AUDIOLOGICAL INTERVENTIONS

Identification of hearing loss in itself has a limited benefit to the patient if no proper rehabilitative measures are taken. In general the provision of hearing aids is an important part of the rehabilitative measures, also for people with an ID. However many people with hearing loss do not use hearing aids. In the general population this is about two third. Also many persons with an ID, who are known to be hearing impaired, do not own hearing aids, or do not use them.[28,29,18] This might be due to certain client-related factors such as lack of understanding, insufficient cooperation or behavioural problems, but also to health care related factors. However some reports were published on successful hearing aid fitting in persons with an ID. Most publications however concern syndrome related case-histories, usually on children as in the Coffin-Lowry syndrome,[30] the del (6q) syndrome,[31] and the Pallister Killian syndrome.[32] Reports on somewhat larger groups have been published by Evenhuis et al,[33,34] Sakai et al,[35] and Nuijten et al.[36]

In 1993, Evenhuis et al reported on a pilot study in twelve middle-aged persons with Down syndrome in which hearing impairment was treated with ventilation tubes (n = 3) and hearing aids (n = 9) in combination with individual habituation training during several months. Two subjects did not accept the hearing aids in spite of a prolonged and gradual training. Subtle positive reactions were reported by staff in the other ten subjects, although no significant improvement could be measured by means of objective methods. These reactions concerned e.g. increased alertness and increased reactions to noise, speech, and music. No change in behavioural problems due to the treatment was observed. The authors concluded that treatment of hearing loss in the adult population with Down syndrome is as such no guarantee for a detectable improvement of communication. They recommended that the influence of several personal and environmental factors had to be studied further.

After hearing assessment in an institutionalised elderly population with a mild to severe intellectual disability by other causes than Down syndrome, twenty four subjects were fitted with hearing aids.[34] In four subjects, hearing aids had to be abandoned because of refusal, carelessness and repeated loss, this in spite of a prolonged habituation training. The other twenty subjects were successfully fitted with one or two hearing aids. During follow-up, hearing aids had to be discontinued in four subjects because of severe physical illness or advanced dementia.

In the study of Sakai et al,[35] thirteen children with Cornelia de Lange Syndrome (CDLS) were fitted with hearing aids in combination with early auditory training. Eight children did not tolerate the devices and kept pulling them out, which is a general experience in persons with CDLS. Hearing aids were successful worn by five subjects, which was demonstrated in three subjects with a moderate to severe hearing impairment by improved sound localisation, as well as by improved discrimination of environmental sound. The other two subjects continued to use their hearing aids, although they did not exhibit any auditory behaviour.

Due to malformations of the ear or recurrent middle ear infections, conventional hearing aids are not always feasible. In such cases, bone conduction hearing aids (BCHA) or bone-anchored hearing aids (BAHA) may be an option. This was reported by Nuijten et al[36] in a group of fifteen persons (mostly children) with a 18q deletion and hearing impairment (partly) as a result of congenital aural atresia. Apart from other ENT-treatment, nine conventional hearing aids were prescribed (eight successful, one not frequently used), two BCHA (one successful, one not frequently used) and one BAHA (successful). The hearing aids resulted in improved auditory reactions, and improved pronunciation of words already known.

In the population with Down syndrome, conventional hearing aids may not be feasible because of the chronic middle ear problems.[37,38] Sheehan & Hans[39] showed that BAHA can be a valuable amplification system in persons with Down syndrome. Because of the complications reported (early complications in 20 out of 43 patients, such as infection or breakdown, skin overgrowth, and failure of osseo-integration), BAHA should be considered only after conventional hearing aids and/or ventilation tubes have failed or are not feasible at all. In spite of the complications, the survey showed a high patient and carer satisfaction.

In the aforementioned studies of Evenhuis[33,34] and Sakai,[35] rehabilitation consisted of hearing aids and individual auditory training. Based on the experience with hearing aid fitting in twelve adults with Down syndrome, Buchanan et al[40] stressed the need of a comprehensive rehabilitation programme in the management of hearing disorder in adults with Down syndrome. This programme should consist not only of habituation training of the hearing impaired person, but also of in-service training of staff on the effect of hearing loss on communication skills, and the benefits and limitations of amplification, further on hearing instrument usage, care and troubleshooting. In addition frequent otologic check-ups should take place because of the frequently occurring middle ear problems in people with Down syndrome. In case of self-injurious behaviour a consistent programme of behavioural management should also be provided.

Many persons with an ID by other cause than Down syndrome are also dependent on carers or may exhibit self-injurious behaviour. So the offering of such a programme should in our opinion not be limited to the population with Down syndrome.

However if we focus on the hearing aids themselves as part of the rehabilitation programme, we do not know whether adults with an ID have the same expectations of hearing aids as adults from the general population, or whether their experiences with the hearing aids afterwards are comparable with other adults. If not, this might have consequences for the introduction of hearing aids, the fitting, and the follow-up. Thus far, no research results have been published on these issues. Therefore a pilot study was established in a group of sixteen adults with a mild or moderate ID.

Aims of the hearing aid study

Aims of this study in adults with an ID were to explore before the fitting the attitude and expectations towards hearing aids, and afterwards to identify the elements that contribute to satisfaction (**chapter 4**).

AUDIOLOGICAL CARE IN THE NETHERLANDS FOR THE POPULATION WITH AN INTELLECTUAL DISABILITY

In the Netherlands, the necessary technological expertise and equipment to diagnose and treat hearing impairment in difficult-to-test clients is available on a district level and covered by the health insurance system. Audiological care for persons with a mild or moderate ID, living in the community, is normally provided by Ear Nose Throat (ENT) specialists. People with lower levels of functioning, behavioural problems or multiple handicaps are now increasingly being referred to district audiological centres, if necessary after ENT treatment of middle ear pathology. In contrast with the situation in several other countries, Dutch audiologists are usually physicists, with a four-year post-doc training in audiology. They provide technical diagnostics and rehabilitation, including hearing aid fitting. Individual auditory training and family guidance is only offered to young children and their family if a hearing loss of 70 dB or over has been diagnosed. In case of multiple handicaps, also children with a moderate hearing impairment can be referred. For adults with an ID, such guidance programmes do not exist. On request, as an extra service, audiological centres may organise a one-time informative meeting for staff.

PROTOCOL FOR AUDIOLOGICAL REHABILITATION

After the development of the Dutch guidelines for early identification and diagnosis of hearing impairment in persons with an ID in 1999 a Dutch working group made an inventory of barriers in the audiological care of people with an ID. The team was chaired by Verschuure and Evenhuis, and consisted of representatives from audiological centres, specialised centres for deaf and hearing-impaired people, and ID centres. Based on the identified barriers, a four module protocol was developed for audiological rehabilitation in persons with an ID (**Chapter 5**). In the protocol several recommendations of the aforementioned rehabilitation programme of Buchanan can be recognised. However one important issue was missing in Buchanan's programme: assessment of acoustical conditions in ID services. Living rooms in institutes, in community based residences, as well as in day-care centres, often are larger than living rooms in ordinary houses. Also surfaces of walls, floor and furniture usually are smooth because they have to be easy to clean, and to be suitable for intensive wheelchair use. Due to this, reverberation time increases, which increases the level of background noise. This poses a problem to persons with a sensorineural hearing loss whose speech intelligibility is diminished in noisy surroundings. Hearing aids will amplify this noise too. Bad acoustical conditions may therefore on its own cause a failure in hearing aid acceptance. However no guidelines existed for optimal acoustical conditions in living rooms of ID services. Therefore, first guidelines were developed for acoustics in group homes and day-care centres by Verschuure (Dept ENT/Audiology, Erasmus Medical Center Rotterdam) in collaboration with Nijs and Van Berlo (Faculty of Architecture, Technical University Delft), based on existing knowledge on the required threshold signal-to-noise ratios for hearing-impaired people[41] and on the relationship between speech intelligibility and room acoustics.[42]

IMPLEMENTATION OF THE REHABILITATION PROTOCOL

After the development of the audiological rehabilitation protocol our next step was to investigate the feasibility of the implementation of this protocol in ID services. Just spreading this protocol in ID services and expecting that disciplines involved will adopt them and change their professional behaviour according to the protocol was considered insufficient. A more active approach was chosen. It is shown in medical practice that the implementation of any innovation should be considered in a holistic, contextual manner, as many factors from different levels may play a role in the adoption of the innovation.[43] It is therefore essential to identify determinants in care organisations that may facilitate or impede the implementation process,[44] with subsequent development of strategies to overcome them.

After a careful preparation, taking the aforementioned into account, a prospective descriptive study was established in the year 2000 to investigate the feasibility of the implementation of this protocol in residential, as well as in community-based homes, and day-care centres for people with an ID. In the second part of this study the implementation process was evaluated, and factors identified that influenced the implementation process.

Aims of the implementation study

Aims of the first part of the implementation study were to study the feasibility of adequate audiological rehabilitation in ID services, to describe obstacles that would be met in the implementation process, and to describe initiatives that facilitated the implementation process (**chapter 5**).

Aim of the second part of the implementation study was to determine factors that influence the implementation of audiological rehabilitation in ID services at an organisational level (**chapter 6**).

The last chapter of this thesis (**chapter 7**) presents a comment on our findings and the problems that were met during the study. In addition, suggestions for future research will be offered, and the consequences of the study results discussed.

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2 Chapter

PREVALENCE OF HEARING IMPAIRMENT IN 1598 ADULTS WITH AN INTELLECTUAL DISABILITY: CROSS-SECTIONAL POPULATION BASED STUDY

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ABSTRACT

A cross-sectional epidemiological study on hearing impairment was carried out in an age-Down syndrome-stratified random sample of 1598 persons drawn from a base population of 9012 persons, representative for the Dutch adult population of intellectual disability (ID) service users. The re-weighted population prevalence is 30.3% (95% confidence interval [CI]: 27.7 - 33.0%). Subgroup prevalences range from 7.5% (95% confidence interval [CI]: 3.6 - 13.3) in the subgroup aged 18 - 30 years with ID by other causes than Down syndrome, up to 100% (95% CI: 79.4 - 100%) in adults over 60 years of age with Down syndrome. Down syndrome (OR: 5.18, 95% CI: 3.80 - 7.07) and age were confirmed to be risk factors. Age-related increase in prevalence in persons with Down syndrome appears to occur approximately three decades earlier, and in persons with ID by other causes approximately one decade earlier than in the general population.

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1 Introduction

At the end of the last century, several large epidemiological studies were undertaken to estimate prevalence of hearing impairment in general adult populations. These studies, performed in Great Britain, Italy and Australia, all show prevalences of 16 - 17%,[1,2,3] with hearing impairment defined as a loss of 25 dB and over, averaged over the frequencies 0.5, 1, 2, and 4 kHz. We wondered whether prevalences in the population with an intellectual disability (ID) would be comparable or higher. It is for instance known that people with Down syndrome are at risk of an early onset of age-related hearing impairment.[4,5] Further, as a result of improved health care for people with an ID, their life expectancy increases. Herewith the number of people with ID that develop age-related hearing impairment is also expected to increase. So far no population-based studies on this topic specifically in people with an ID have been published.

In order to provide epidemiological data, a large-scale population-based, cross-sectional study of adult users of ID services was set up in the Netherlands. Apart from the prevalence of hearing impairment, also the prevalence of visual impairment was studied.[6] The following research questions were formulated:

1. What is the prevalence and severity of hearing impairment in adults with ID?
2. In how many cases had hearing impairment not been diagnosed prior to the study?
3. How is the prevalence of hearing impairment related to age, Down syndrome and severity of ID?

2 POPULATION AND METHODS

2.1 Population

Fourteen ID services, distributed over the Netherlands, with a total base population consisting of 9012 clients aged 18 years and over, consented to participate. Participating ID services were - as to the number of clients they represented - almost equally divided between residential and community-based services. This represented the situation in the Netherlands at the start of the study in 1998. Given our resources in money and manpower, assessment of a total sample size of 2100 was considered feasible. Because old age and the presence of Down syndrome are known risk factors for hearing impairment, testing of differences in prevalence of visual and hearing impairment between four subgroups (50+/- and Down syndrome +/-) was incorporated in the design of the study. From the distribution of age and presence of Down syndrome in the base population, and provisional figures available from an assessment-based study of sensory impairments in institutionalised adults which was in progress in the Netherlands,[7] it was inferred that a non-stratified random sample would yield an unnecessarily large subgroup of young persons without Down syndrome (1260) and a subgroup of people with Down syndrome aged 50 years and over (84) that was too small for statistical analysis. Therefore an age-Down syndrome-stratified random sample with 1000 persons in the young group without Down syndrome and 200 persons in the older group with

Down syndrome was taken. This was done because the latter is a vulnerable group, with a high prevalence of dementia and early death. It was feared that, in this group specifically, a relatively large number of participants might be lost between consent and completion of all measurements. The remaining two groups were slightly over-sampled. In this way a sufficient number of persons in each subgroup to detect differences between groups with a power of 0.80 and $\alpha = 0.05$ was secured.

After approval of the Medical Ethical Committee, written informed consent was obtained from ID service-providers, participants and/or their legal representatives. If a selected client had moved or died before the consent procedure was completed, or if no consent was obtained, he or she was randomly replaced by a client from the same subgroup.

2.2 Methods

In a flow chart (figure 1) the screening protocol is shown. The medical records were checked for cause and degree of ID and for audiometry data. If reliable audiometry data of two years old or less were available, no further screening or audiometry was performed. After removal of earwax by the ID physician or general practitioner, on-site screening of hearing function took place in the ID services by audiologists in-training or audiology-assistants from two specialised organisations for the deaf and hearing impaired. Persons who failed the screening were assessed on site by two E.N.T. specialists who were part of the research group, and treated if necessary, before referral to a regional audiological centre.

Ten district audiological centres were involved in the audiometric assessments, which were performed on site or in the audiological centres. Audiologists were personally informed about the study and the audiometry protocol, as well as during meetings of the Federation of Dutch Audiological Centres.

2.3 Definitions and protocol

Degree of intellectual disability was classified as mild for IQ 55 - 70, moderate for IQ 35 - 55, severe for IQ 25 - 35, and profound for IQ below 25.

Screening was performed with Distortion Product Oto Acoustic Emissions (DP-OAE) and Transient Evoked Oto Acoustic Emissions (TE-OAE).[8,9]

Audiometry consisted of:

1. Pure Tone Audiometry and if possible speech audiometry, if necessary using conditioning methods (play audiometry). Measurement of air conduction and if possible of bone conduction thresholds at 0.5, 1, 2 and 4 kHz, if masking was accepted.
2. Auditory brainstem response (ABR) audiometry, if necessary after oral sedation, in clients who could insufficiently cooperate with pure tone audiometry. For sedation, additional consent of legal representatives was asked.

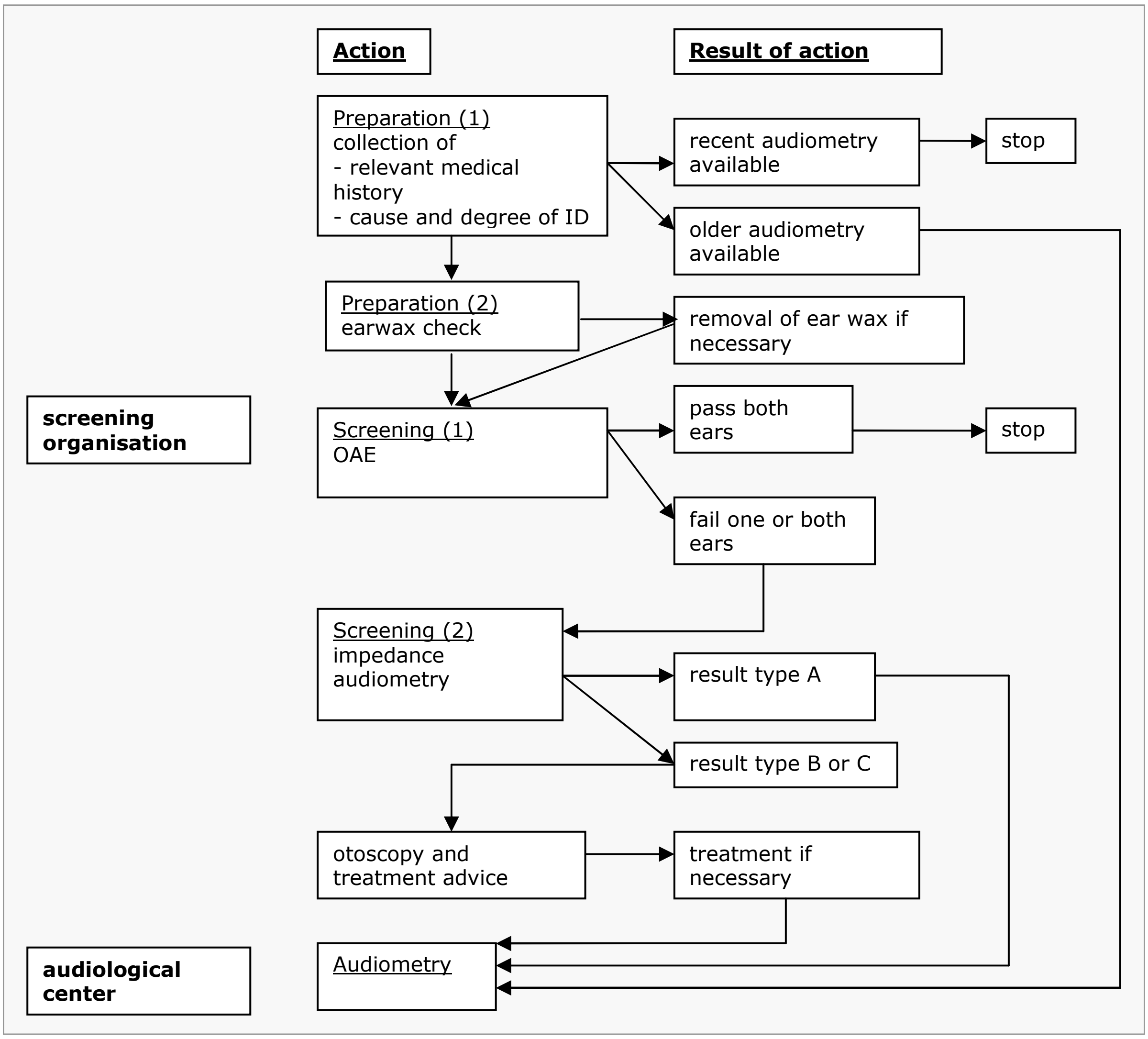

Figure 1 *Preparations, screening and audiometry*

Hearing impairment was defined, according to the criteria of the World Health Organization of 1997[10] as an average loss of more than 25 dB at the best ear. The degree of hearing impairment was classified as a mild impairment for losses between 26 - 40 dB, moderate impairment for losses between 41 - 60 dB, severe impairment for losses between 61 - 80 dB, and as profound impairment for losses over 80 dB.[10] However, we did not average this loss over four frequencies according to the WHO definition, but over three (1, 2 and 4 kHz), according to the International Consensus Statement of the International Association for the Scientific Study of Intellectual Disabilities.[11]

To estimate a hearing threshold based on ABR audiometry, 10 dB was deducted from the response threshold.[12] The result of the ABR audiometry was also classified according to the WHO criteria, because the hearing threshold at 3 kHz is crucial for speech recognition in noise[13] and has a one-to-one relationship with the pure tone threshold in cochlear hearing loss.[12]

2.4 Analysis

All data were stored in Access 97 (Microsoft Corporation, Redmond, USA) and analysed with SPSS version 11.0 (SPSS Inc., Chicago, Illinois, USA). Multiple logistic regression was used to evaluate the independent relationships between hearing impairment on the one hand, and Down syndrome, degree of ID and age 50-/50+ on the other hand.

We generalised the prevalence of hearing impairment in the study population to the prevalence in the base-population of 9012 participants (which was representative for the Dutch adult ID population) as follows: to compensate for participants with missing audiometry test and for the stratified sample (age and Down syndrome), a re-weighting technique was applied.[14] This technique estimates the probability that a participant is included in the study population with non-missing audiometry, based on prognostic variables (age, Down syndrome, degree of ID, gender, residential/community and the interaction between Down syndrome and degree of ID). A re-weighted prevalence is computed by weighing each patient proportionally to the inverse of the above-mentioned probability. After this, a weighted prevalence was computed from the figures found in the 2 x 2 x 2 cells: age 50-/50+, Down syndrome -/+, residential/community care (direct standardisation).

We wanted to compare our study results with the results of the three aforementioned epidemiological studies in general populations. However in these studies, hearing impairment was defined as a loss of 25 dB and over in the better ear, averaged over four frequencies (0.5, 1, 2 and 4 kHz), instead of our definition of more than 25 dB, averaged over three frequencies (1, 2 and 4 kHz).

In order to consider what difference this would yield for prevalence numbers, we estimated prevalences of hearing impairment as a loss of more than 25 dB, averaged over three (1, 2 and 4 kHz) as well as over four frequencies (0.5, 1, 2 and 4 kHz) in all participants with reliably measured hearing thresholds in four frequencies. The only remaining difference then would be that we did not include hearing losses of exactly 25 dB in the diagnosis of hearing impairment. If we had done so, resulting prevalences might have been slightly higher.

3 RESULTS

3.1 Population

A detailed account of consent, inclusion and participation has been published elsewhere.[15] A random sample of 2706 participants was approached, consent for participation was obtained from 1598. For 634 participants of the 996 for whom no consent was given, the distribution of the housing situations was almost the same as the distribution for the ID population in the Netherlands. The other 362, for whom consent was not obtained because of logistic problems, all lived in the community, leading to an overrepresentation of more severe ID in the study group. The final study population of 1598 persons consisted of 893 males and 705 females. The composition of the study population, according to age, degree of ID and Down syndrome is shown in table 1. Mean age was 45.68 years (range 20.19 - 88.73) in all participants and 45.42 years (range 20.85 - 75.93) in participants with Down syndrome.

Table 1 Composition of the study population (n = 1598) (percentages in parentheses)

Age	**Down -** %	**Down +** %	**Total** %
Subjects < 50 years			
Mild ID (IQ 55 - 70)	161 (10.1)	23 (1.4)	184 (11.5)
Moderate ID (IQ 35 - 55)	270 (16.9)	145 (9.1)	415 (26.0)
Severe ID (IQ 25 - 35)	147 (9.2)	55 (3.4)	202 (12.6)
Profound ID (IQ < 25)	108 (6.8)	19 (1.2)	127 (7.9)
Unknown	58 (3.6)	15 (1.0)	73 (4.6)
Total	744 (46.6)	257 (16.1)	1001 (62.6)
Subjects ≥ 50 years			
Mild ID (IQ 55 - 70)	82 (5.1)	7 (0.4)	89 (5.6)
Moderate ID (IQ 35 - 55)	179 (11.2)	74 (4.6)	253 (15.8)
Severe ID (IQ 25 - 35)	84 (5.3)	45 (2.8)	129 (8.1)
Profound ID (IQ < 25)	43 (2.7)	25 (1.6)	68 (4.3)
Unknown	46 (2.9)	12 (0.8)	58 (3.6)
Total	434 (27.2)	163 (10.2)	597 (37.4)

ID	*intellectual disability*
Down -	*intellectual disability by other causes than Down syndrome*
Down +	*Down syndrome present*

3.2 Participation and cooperation

In table 2, results are shown of participation in the subsequent phases of the assessment protocol.

For 1215 out of the 1598 participants reliable data could be obtained: 242 because of recently completed audiometry, 353 who passed the screening at both ears, 31 who passed the screening at one ear (and thus by definition were not classified as hearing impaired), and 589 in whom interpretable audiometry data were obtained.

For the 383 participants in whom no reliable data could be obtained, distribution of ID is shown in table 3. More severe and profound ID appear to be present in this group, and in both subpopulations with Down syndrome and with ID by other causes, as compared to the whole study population. Twenty-six participants withdrew before the start of the screening because of death (n = 7), moving to another location (n = 4), consent withdrawn (n = 3), consent for file inspection only (n = 6), age-related decline (n = 1), no reasons noted (n = 5). In 113 participants the OAE screening failed because of uncooperative behaviour. Screening was not performed in 56 participants because they (repeatedly) did not show up at the appointment. In 235 clients who failed the screening, audiometry had not been performed because of death (n = 16), moving to another location (n = 3), dementia (n = 5), no permission for necessary sedation for ABR audiometry (n = 28), no show at the appointment (n = 6), logistical reasons (n = 25) and reasons unknown (n = 152). In 37 of these 235 participants, hearing impairment had been diagnosed longer than two years ago. Because of the sensorineural component in the hearing impairment in 17/37 participants, it could be established with certainty that they would still be hearing impaired. However, classification of the severity in the present situation was not possible.

In 122 cases, participants were not testable (n = 48) or results uninterpretable (n = 74).

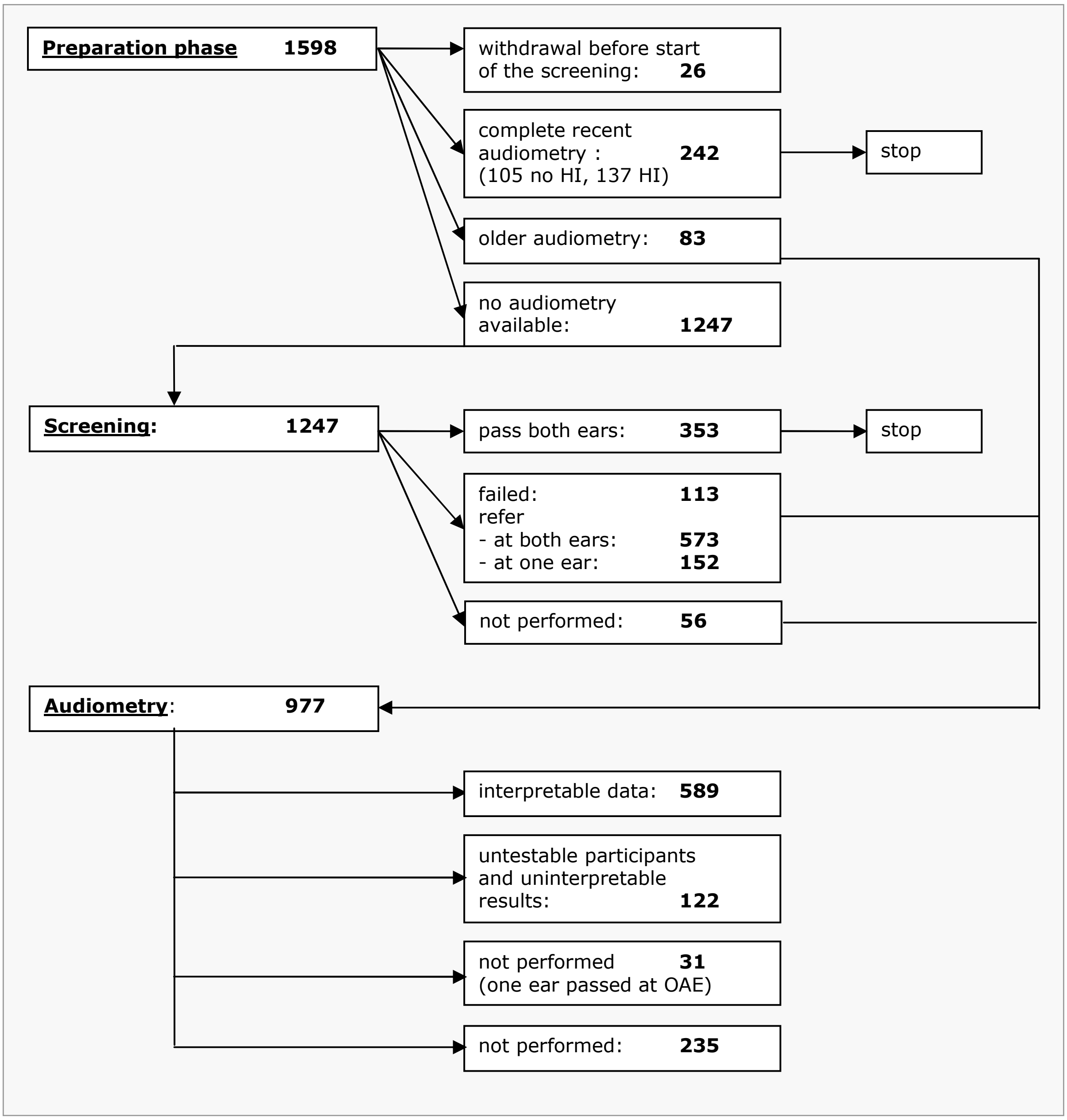

Table 2 Results of preparation, screening and audiometry (n = 1598)

HI hearing impairment

Table 3 Distribution of ID and Down syndrome in the total study population and in the group of 383 persons in whom determination of hearing function was not possible (absolute numbers in parentheses)

	mild and moderate ID %	**severe and profound ID %**	**unknown %**
Whole study population (n = 1598)	58.9 (941)	32.9 (526)	8.2 (131)
Down syndrome present	15.6 (249)	9.0 (144)	1.7 (27)
ID by different causes	43.3 (692)	23.9 (382)	6.5 (104)
No determination of hearing function (n = 383)	43.6 (167)	47.0 (180)	9.4 (36)
Down syndrome present	11.5 (44)	15.1 (58)	1.8 (7)
ID by different causes	32.1 (123)	31.9 (122)	7.6 (29)

ID *intellectual disability*

3.3 Deviations from the protocol

In 21/651 participants (including participants with recently completed audiometry) in whom pure tone audiometry was performed, not all hearing thresholds at 1, 2 and 4 kHz were available in both ears in order to calculate an average over the three frequencies. For these persons, the research team individually examined similarity of hearing profiles of the left and right ear and the tendency of hearing thresholds measured at the different frequencies. In this way, hearing impairment could be excluded or diagnosed and classified in 20/21 participants. In one participant the results were uninterpretable.

It appeared that the diagnostic protocol was not applied by audiological centres in all referred participants. In 198 cases, behavioural audiometry was used, instead of Pure Tone Audiometry or ABR audiometry (which was only used in 15 cases). Audiologists reported that ABR audiometry had not been tried because of reluctance to sedation, which was often necessary, a preference for behavioural audiometry, or anticipated difficulties to interpret ABR audiometry results in this group. Because in behavioural audiometry, interpretation of results is based on individual and local circumstances (e.g. degree of ID, alertness and cooperation of client, environmental noise, acoustical conditions) diagnosticians were asked to classify the results themselves, according to the WHO criteria. If results could only be classified in a wider range, the best result was used in the analysis (= lowest degree of hearing impairment). In 34 of the 198 cases, the behavioural audiometry results were not interpretable.

3.4 Prevalence of hearing impairment in the study population

In 424 of the 1215 participants (34.9%) in whom determination of hearing function was possible, hearing impairment was diagnosed. This had not been known prior to the study in 202/424 participants (47.6%). With the inclusion of 17 participants with a diagnosis of permanent hearing impairment, based on older audiometry results, prevalence of hearing loss became 35.8% (441/1232).

3.5 Severity of hearing impairment

Table 4 shows the relationship between a more severe hearing impairment (moderate-profound) and age and Down syndrome. Hearing impairment was significantly more severe in persons aged 50 years and over than in persons younger than 50 years, both in the total study population and the subpopulation with Down syndrome. In the subpopulation with ID by other causes than Down syndrome, this relationship was just not significant (p = 0.07).

Table 4 Moderate-profound hearing impairment in relation to age and Down syndrome (absolute numbers in parentheses)

	> 40 dB	**95% CI**
hearing impaired (n = 424)	51.4% (218)	
< 50 years	41.1% (79)	34.1 - 48.5%
Down -	45.5% (40)	34.8 - 56.4%
Down +	37.5% (39)	28.2 - 47.5%
≥ 50 years	59.9% (139)	53.3 - 66.3%
Down -	58.6% (85)	50.2 - 66.7%
Down +	62.1% (54)	51.0 - 72.3%

Down -	*intellectual disability by other causes than Down syndrome*
Down +	*Down syndrome present*

3.6 Re-weighted prevalences in the total population of adult ID service users

The re-weighted prevalence of hearing impairment was 30.3% (95% CI: 27.7 - 33.0%) in the representative base population of 9012 adults with an ID. In the subpopulation with Down syndrome a re-weighted prevalence of 57.4% (95% CI: 51.6 - 62.9%) could be estimated, and in the subpopulation with ID by other causes of 24.2% (95% CI: 21.4 - 27.1%). The distribution of mild versus more severe HI was around 50 - 50% in the total population, as well as in the subpopulations with Down syndrome and with ID by other causes (table 5).

Table 5 Re-weighted prevalences of degree of hearing impairment in the total population of ID service users, and in subpopulations with Down syndrome and ID by different cause

	26 - 40 dB	**95% CI**	**> 40 dB**	**95% CI**
Total population	14.9%	13.0 - 17.0%	14.5%	12.6 - 16.6%
Subpopulation with Down syndrome	31.1%	26.1 - 36.7%	26.0%	21.3 - 31.3%
Subpopulation with ID by a different cause	11.2%	9.1 - 13.3%	11.8%	9.8 - 14.2%

3.7 Relationship of hearing impairment to Down syndrome, age, and degree of ID (table 6)

3.7.1 Prevalence and Down syndrome

In people with Down syndrome, both under and over 50 years, prevalence numbers were much higher than in the rest of the study population. The effect of Down syndrome as compared to persons with ID by other causes, irrespective of age, was assessed by a multiple logistic regression analysis with hearing impairment as the dependent variable: the odds ratio of hearing impairment appeared 5.18 (95%: CI 3.80 - 7.07, data not shown).

Table 6 Prevalence (%) of hearing impairment by age, degree of ID and Down syndrome (n = 1215) (absolute numbers in parentheses)

	Down – (n = 88)	**95% CI**	**Down +** (n = 104)	**95% CI**
Subjects < 50 yrs				
Mild ID	10.6	5.9 - 17.2	40.0	19.1 - 63.9
Moderate ID	13.6	9.4 - 18.7	47.5	38.4 - 56.8
Severe ID	20.7	13.7 - 29.2	60.0	42.1 - 76.1
Profound ID	19.0	10.2 - 30.9	90.0	55.5 - 99.7
Unknown	15.6	6.4 - 29.5	72.7	39.0 - 94.0
Total	15.1	12.2 - 18.2	52.5	45.3 - 59.6
	Down – (n = 145)	**95% CI**	**Down +** (n = 87)	**95% CI**
Subjects ≥ 50 yrs				
Mild ID	49.2	36.1 - 62.3	50.0	6.8 - 93.2
Moderate ID	45.9	37.7 - 54.3	79.7	67.2 - 89.0
Severe ID	44.6	31.3 - 58.5	72.4	52.8 - 87.3
Profound ID	36.0	18.0 - 57.5	83.3	51.6 - 97.9
Unknown	43.3	25.5 - 62.6	77.8	2.8 - 60.0
Total	45.3	39.8 - 50.9	77.0	68.1 - 84.4

ID	*intellectual disability*
Down -	*intellectual disability by other causes than Down syndrome*
Down +	*Down syndrome present*

3.7.2 Prevalence and age

Age ≥ 50 years was found to increase prevalence numbers in the subgroup with ID by other causes than Down syndrome (table 6). Multiple logistic regression analysis with hearing impairment as the dependent variable (table 7) confirms the effect of age of 50 years and over, both in groups with Down syndrome and with ID by other causes. Figure 2 shows a steady rise of prevalences from age < 30 years onwards in both subgroups.

Table 7 Summary of a multiple regression model with hearing impairment as the dependent variable

		Odds Ratio	95% Confidence Interval	p-value
Down syndrome absent				
Degree of ID*:	mild***	-	-	-
	moderate	1.08	0.70 - 1.66	0.73
	severe	1.38	0.83 - 2.27	0.21
	profound	1.13	0.61 - 2.13	0.70
	missing	1.09	0.57 - 2.08	0.80
Age 50+ (50- reference category)		4.71	3.43 - 6.47	0.00
Down syndrome present				
Degree of ID**:	mild***	-	-	-
	moderate	1.69	0.70 - 4.09	0.24
	severe	2.10	0.78 - 5.63	0.14
	profound	6.66	1.50 - 29.50	0.01
	missing	3.38	0.90 - 12.72	0.07
Age 50+ (50- reference category)		2.73	1.61 - 4.64	0.00

* *test for trend: p = 0.35, category 'missing' excluded*

** *test for trend: p = 0.01, category 'missing' excluded*

*** *reference category*

3.7.3 *Prevalence and degree of ID*

In most subgroups, no significant effect of the degree of ID was demonstrated by multiple logistic regression (table 7). A test for trend showed a significant relationship in participants with Down syndrome (p = 0.01), but not in persons with ID by other causes than Down syndrome (p = 0.35).

3.8 Comparison with prevalences in general adult populations

3.8.1 *Effects of different criteria for hearing impairment on prevalences*

We estimated prevalences of hearing impairment, averaged over three and four frequencies, in 778 people from our database, in whom reliable thresholds were available for 0.5, 1, 2 and 4 kHz. These were identical: 54.4% for three, and 54.5% for four frequencies.

3.8.2 *Comparison with general population figures*

In table 8 and figure 2, age-related prevalences in subpopulations with Down syndrome and with ID by different cause are compared with published prevalence figures in comparable age groups in adult general populations.[1,2] In both ID subpopulations, prevalence is higher than in the general population in all age groups. As compared with the study of Davis,[1] there are no overlapping 95% confidence intervals. In the subpopulation with Down syndrome, age-related rises in prevalence figures occur approximately three decades earlier than in the British and Italian study, and in the subpopulation with an ID by different causes, approximately one decade earlier (figure 2).

Table 8 Comparison of prevalences in the subgroups with and without Down syndrome with general adult population studies in Great Britain[1] and Italy.[2]

Age group (years)	**Down syndrome present***	**95% CI**	**Down syndrome absent***	**95% CI**	**British study****	**95% CI**	**Italian study****
18 - 30	36.4	20.4 - 54.9	7.5	3.6 - 13.3	1.8	0.7 - 2.9	1.9
31 - 40	46.2	34.8 - 57.8	12.0	8.0 - 17.1	2.8	1.2 - 4.4	3.9
41 - 50	64.4	53.4 - 74.4	22.3	17.1 - 28.2	8.2	6.1 - 10.3	8.3
51 - 60	73.7	63.6 - 82.2	32.0	25.3 - 39.4	18.9	16.1 - 21.7	18.7
61 - 70	100	79.4 - 100	52.3	41.3 - 63.2	36.8	32.4 - 41.2	37.7
71 - 80	numbers too small		79.2	65.9 - 89.2	60.2	53.0 - 67.5	69.4

* *defined as a hearing impairment > 25 dBHL in the better ear averaged over the frequencies 1, 2 and 4 kHz*

** *defined as a hearing impairment ≥ 25 dBHL in the better ear averaged over the frequencies 0.5, 1, 2 and 4 kHz*

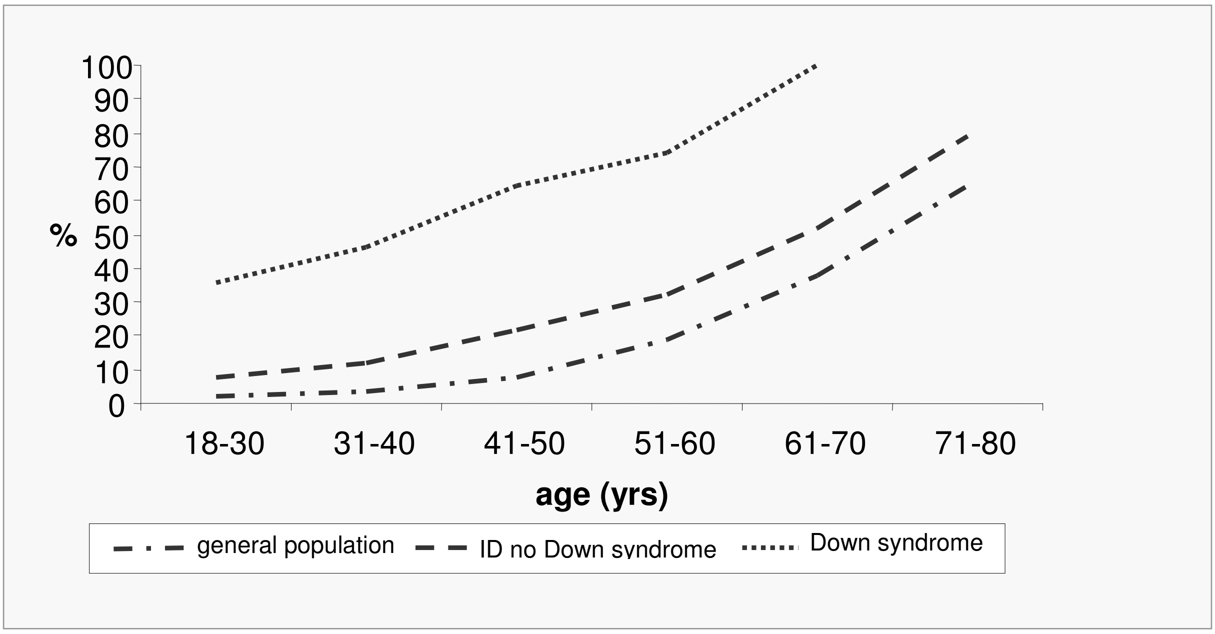

Figure 2 *Hearing impairment in age groups*

4 DISCUSSION

This first nationwide epidemiological study of hearing impairment in adult users of intellectual disability (ID) services shows, that with a re-weighted population prevalence of 30.3% (95% CI: 27.7 - 33.0%), hearing impairment occurs in almost one out of three adults with an ID, which is two times higher than the prevalence of 16 - 17% in general populations.[1,2,3] This hearing impairment had not been diagnosed prior to this study in almost half of the cases (47.6%), which implies that many cases of hearing impairment will remain undiagnosed in ID services where no hearing screening takes place. This high prevalence is not only accounted for by the very high re-weighted prevalence of 57.4% in adults with Down syndrome, but also by the significantly increased prevalence of 24.2% (95% CI: 21.4 - 27.1%) in adults with an ID by other causes, as compared to the aforementioned general adult populations. We showed that the differences could not be explained by the use of slightly different definitions of hearing impairment.

Incomplete inclusion in a large community-based setting because of logistical reasons, skewed the study population towards more severe degrees of ID. However, the computation of weighted prevalences, taking into account type of care, age and Down syndrome, compensated for the bias that was created by stratification and for any bias in the prevalence estimate due to non-response dependent on these three characteristics. Because no medical information was available from those who did not consent, skewing towards more or less seriously affected participants (e.g. multiple handicaps or behavioural problems) cannot be ruled out.

It needs to be noted that the studied population consisted of ID service users only (homes, day-activity centers, supported living), so the present figures are not representative for unregistered people. In the Netherlands, these are primarily persons with mild or borderline ID, who may report hearing problems themselves.

Previously published data on hearing impairment in persons with ID have been based on questionnaires,[16,17] on small scale community-based studies,[18,19] or on studies in selected groups.[20,7] These studies resulted in prevalences varying from 12 - 47%. This broad range however was also caused by a lack of uniformity in definition of hearing impairment.

Apart from the prevalence, also the severity of hearing impairment appears to be increased in adults with an ID. In the general population.[1,2,3] mild hearing impairment, defined as losses of 25 - 45 dB (12.2% - 13.8%) is about four times more frequent than moderate to profound hearing impairment (2.8 - 4.0%). In the adult population with ID, the distribution is equal (14.9% and 14.5% respectively). This also applies to subpopulations with Down syndrome (31.1% and 26.0% respectively), and with an ID by different causes (11.2% and 11.8% respectively). It is not very likely that the entire difference is explained by our inclusion of losses of 41 - 45 dB into moderate hearing impairment.

Down syndrome and age were confirmed to be risk factors, as was already suggested by previous studies[4,7] but the relationship could, for the first time, be validly quantified in the present study. Hearing impairment occurs more frequently in the subpopulation with Down syndrome (odds ratio 5.18), as compared with the subpopulation with ID by other causes. In adults with Down syndrome, hearing impairment may have a conductive and/or sensorineural nature. The conductive component may be caused by congenital anomalies of the middle ear, e.g. malformation of the ossicles,[21,22] or by the frequently occurring or chronic middle ear infections,[23,24] persisting into adulthood.[5] These may secondarily lead to chronic perforation of the tympanic membrane or cholesteatoma,[25] but also to sensorineural hearing loss.[26,27] In addition to this, sensorineural hearing loss may also be caused by ototoxic medication that is prescribed because of the infections. Age-related sensorineural hearing impairment in Down syndrome, which already appears during the second decade of life, shows similar characteristics as presbyacusis.[4,5]

The influence of age in the group 50 years and over is stronger in the subpopulation with ID by other causes (odds ratio 4.71), than in the subpopulation with Down syndrome (odds ratio 2.73) (table 7). This may have been caused by the already high prevalences in the subpopulation with Down syndrome younger than 50 years. A combination of these two risk factors resulted in a prevalence of over 70% in the subpopulation with Down syndrome, older than 50 years, reaching 100% over age 60 years (95% CI: 79.4% - 100%) (table 8). This implies that persons with Down syndrome, aged 50 years and over, should be considered hearing impaired until proven otherwise.

The risk of hearing impairment appeared to be significantly increased in all 10-year age groups from 18 - 30 years onwards, as compared to the general population (table 8, figure 2). This was already shown for the subpopulation with Down syndrome by Buchanan,[4] (1990), but it also applies to the subpopulation with an ID by different cause. As a result, in the latter subpopulation, age-related increase of hearing impairment appears to occur a decade earlier than in the general population. We do however not a priori expect premature ageing on top of the subliminal congenital or early childhood impairments in this group. Conductive losses might partially explain the increased risk.

Influence of degree of ID on prevalence of hearing impairment could not be demonstrated with multiple logistic regression analysis, but could be established in the subpopulation with Down syndrome with a test for trend (p = 0.01). This shows that the effect is less clear than in visual impairment in the same study population, for which severe or profound ID is by far the most important risk factor.[6] A possible explanation for the difference may be the fact, that screening of visual function also detects low visual acuity due to cerebral damage. However with oto-acoustic emissions, which are produced by the outer hair cells in a normally functioning inner ear, no hearing impairment will be detected that has its origin from the inner hair cells onward. Since in 1996, the concept of auditory neuropathy was introduced by Starr et al,[28] referring to a condition of the auditory nerve in which oto-acoustic emissions are preserved, and auditory brainstem potentials are absent or severely distorted, several study results were published on this topic. It has been shown e.g. for the neonatal intensive care population and for children who had hyperbilirubinemia, that they have an increased risk of auditory neuropathy.[29,30,31] It can be hypothesised that if auditory brainstem response screening equipment would be used, the prevalence of hearing impairment, identified in adults with an ID, may even be higher. At present, such equipment is used in neonatal hearing screening, and not available for use in adults. With this screening method, auditory pathways past the inferior colliculus in the brainstem are not tested. However, because of the many alternate auditory pathways leading from the brainstem to the cortex, the auditory system past this level seldom breaks down completely.

CONCLUSIONS

- The prevalence and severity of hearing impairment are significantly increased in the adult Dutch ID population, as compared with general adult population studies. The significant increase in prevalence applies to all age groups, including the youngest (18 - 30 years), with ID by others causes than Down syndrome.
- Hearing impairment had not been recognised in almost half of the cases prior to the screening.
- Age and Down syndrome could be confirmed and quantified as risk factors, with a very high odds ratio for Down syndrome as compared to adults with ID by different causes.
- The effect of the degree of ID on prevalence of hearing impairment is less clear than in visual impairment.
- Age-related rises in prevalence in persons with Down syndrome appear to occur approximately three decades earlier than in the general population, and in persons with ID by other causes than Down syndrome approximately one decade earlier.

Recommendations

We recommend the following adjustments of the existing IASSID consensus guideline for hearing screening in adults with an ID[11]:

1. to shift the screening of age-related losses forward by 10 years (from 50 to 40 years) for persons with an ID by other causes than Down syndrome
2. to perform in persons with Down syndrome complete audiometry instead of hearing screening every 3 years throughout life.

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3 Chapter

EPIDEMIOLOGY OF DUAL SENSORY IMPAIRMENT IN ADULTS WITH INTELLECTUAL DISABILITIES

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ABSTRACT

Background: People with intellectual disabilities (ID) have an increased risk of both visual and hearing impairment, but no epidemiological information is available on the combination. Dual sensory impairment in the general population is primarily a condition of ageing.

Aim: To estimate in a cross-sectional study the prevalence, severity, and treatable causes of dual sensory impairment in adult ID service users, and the relation of prevalence and severity to risk factors.

Methods: Ophthalmologic and audiometric assessments were performed in a random sample of 1598 persons, stratified for age and Down syndrome, from a representative base population of 9012 adult users of Dutch ID services. WHO definitions were applied.

Results: Dual sensory impairment was identified in 77 cases (32/77 Down syndrome). In nine cases (12%) this was known prior to study. Re-weighted population prevalences: total adult ID population 5.0% (95% CI 3.9 - 6.2); < 50 years 2.9% (1.9 - 4.1) (general population < 65 rough estimate < 0.03%); ≥ 50 11.0% (7.9 - 14.7) (general Dutch 65+ population 1.4% (0.8 - 2.0%)).

As risk factors could be identified: a more severe ID ($p < 0.0001$), age ≥ 50 (OR 3.62, $p = 0.000$), Down syndrome (OR 2.37, $p = 0.001$). Treatable ophthalmologic conditions were diagnosed in 44/77 cases (57%). Hearing losses were sensorineural in 29, conductive in 6, mixed in 24, and unclear in 18 cases.

Conclusions: Adults with ID have a considerably increased risk of dual sensory impairment, both before and after age 50 years, as compared to the general population. This remains unidentified in a majority of cases. Part of the underlying conditions are treatable or can be rehabilitated. Physicians involved in medical care of ID people should cooperate to detect and possibly treat (dual) sensory impairments.

Submitted

1 INTRODUCTION

We have recently shown, that Dutch adults with intellectual disabilities (ID) have a 10 times increased risk of low vision, as compared to the Dutch general population aged 55 years and over,[1,2] and a two times increased risk of hearing impairment, as compared to published prevalences in three general adult populations.[3,4,5,6] Risks were increased at all ages, probably as a result of congenital conditions leading to both brain damage and damage of ocular or auditory structures, which are now increasingly of perinatal origine[7] and early ageing (premature cataracts and hearing loss in Down syndrome),[8,9] whereas in older adults, age-related conditions are superposed upon pre-existing conditions. In over 40% of adults with ID, visual impairment or hearing impairment had not been identified prior to our assessments.[1,3] With such significantly increased risks, a combination of visual and hearing impairments can also be expected to occur regularly. If this diagnosis too would remain obscure in a significant number of cases, this might have an extraordinary impact on the independence and social functioning of these people.

To obtain information on prevalence, severity and treatable causes of dual sensory impairment in the adult population with ID, the association with age, severity of ID and Down syndrome, and missed diagnoses, our data were re-analysed.

2 METHODS

2.1 Study design

Population-based, cross-sectional study.

2.2 Study population

Calculation of sample size, method of randomisation, inclusion and participation, and characteristics of the study population have been reported in detail elsewhere.[1,3] From a representative base population of 9012 adult users of 14 Dutch residential and day-care intellectual disability services, a random sample of 2100, stratified for Down syndrome yes/no and age < 50/50+ years, was drawn, of which 1598 persons were actually included. Mean age was 45.7 years (range 20.2 - 88.7), and 893 were males (55.9%). As a result of incomplete inclusion in community based settings, the distribution was unintentionally skewed towards more severe ID.

2.3 Diagnostic methods

Diagnostic methods for visual functions have been reported in detail.[1] Assessments were performed on-site in the ID services, and included visual acuity tests (Snellen's chart, Burghardt's children's chart, Stycar single characters and matching,[10] Cardiff Acuity Cards,[11] Teller Acuity Cards,[12]) visual fields (confrontation), autorefraction or sciascopy, handheld slitlamp biomicroscopy, assessment of strabism and ocular pressure. To ensure reliable visual acuity measurements, investigators were specially trained in a difficult-to-test population, inter-observer agreement was checked in a group with severe ID, and at least two different acuity tests were applied in all cases.

Hearing screening was performed on-site using Distortion Product Oto Acoustic

Emissions (DP-OAE) or Transient Evoked Oto Acoustic Emissions (TE-OAE), and tympanometry. In case of abnormalities, ENT assessment and further audiometry were performed: pure tone audiometry, or auditory brainstem response (ABR) audiometry in clients who could insufficiently cooperate.[3]

2.4 Definitions

Severity of intellectual disability: mild (IQ 55 - 70), moderate (IQ 35 - 55), severe (IQ 25 - 35), profound (IQ < 25).

Low vision and hearing impairment were diagnosed and classified according to WHO-criteria.[13,14] Visual impairment: visual acuity < 0.30, but not < 0.05 and/or visual fields < 30° around central fixation point; blindness: visual acuity < 0.05 and/or visual fields < 10°. Best corrected visual acuity of the best eye was used in the analyses, but if correction was not accepted, we used presenting visual acuity. Hearing impairment: an average loss for 1, 2 and 4 kHz of more than 25 dB at the best ear. To estimate a hearing threshold based on ABR audiometry, 10 dB was deducted from the response threshold.[15] The degree of hearing impairment was classified as mild (26 – 40 dB), moderate (41 – 60 dB), severe (61 – 80 dB), or profound (over 80 dB).[14] Results of ABR audiometry were classified similarly, because the hearing threshold at 3 kHz is crucial for speech recognition in noise[16] and has a one-to-one relationship with the pure tone threshold in cochlear hearing loss.[15]

Refractive error: spherical refractive error > ± 1.00 D and/or cylindrical refractive error > -2.00 D. Severe hypermetropia/myopia: refractive error ≥ ± 5.00 D. Slitlamp binocular biomicroscopy was applied to detect corneal opacities, keratoconus and cataract. Cataract: any opacity of the lens visualised in the physiologically widened pupil in a darkened room. Keratoconus: any obvious conal deformity. Corneal opacities: any opacity of the cornea. Ocular hypertension: ocular pressure ≥ 21 mm Hg.

Type of hearing impairment was established as sensorineural, if the pure tone audiogram showed a mean air-bone gap of less than 7.5 dB and no air-bone gap exceeding 10 dB.[15] If bone conduction could not be assessed, sensorineural hearing loss was diagnosed in case of a history without middle ear pathology, and no signs of middle ear pathology at tympanometry and ENT assessment, whereas conductive hearing loss was diagnosed in case of signs of middle ear pathology at ENT assessment and/or if tympanometry indicated middle ear pathology (type B).[17]

2.5 Analysis

To compensate for participants with incomplete data and for the stratified/skewed sample, re-weighting[18] was applied, in order to obtain valid prevalences for the representative base population of 9012 ID adults. Re-weighted prevalences were also computed for the subgroups age < 50/50+ years and no Down syndrome/Down syndrome. Independent relationships between dual sensory impairment on the one hand and Down syndrome-/+, severity of intellectual disability, and age < 50/50+ years on the other hand, were evaluated by means of multiple logistic regression analysis and a test for trend.

3 RESULTS

3.1 Prevalences and associations

A diagnosis of dual sensory impairment could be made or excluded in 1359/1598 participants. The other 239 cases could not be judged, because both visual and hearing function could not be reliably measured (n = 70), or because visual function (n = 70) or hearing function (n = 99) could not be determined, whereas the other function was impaired. Dual sensory impairment was diagnosed in 77/1359 participants (5.7%). The re-weighted prevalence for the total adult ID population was 5.0% (95% CI: 3.9 – 6.2%).

For different levels of severity, re-weighted prevalences were: 2.1% (95% CI: 1 - 3%) for visual impairment and mild hearing impairment, 1.5% (95% CI: 0.9 - 2%) for visual impairment and moderate to profound hearing impairment, 0.4% (95% CI: 0.1 - 0.9%) for blindness and mild hearing impairment, and 0.5% (95% CI: 0.2 - 1%) for blindness and moderate to profound hearing impairment. As a result of separate re-weighting procedures for the subgroups, these figures do not exactly add up to the total prevalence of 5.0%. Re-weighted prevalences by age < 50/50+ years and no Down syndrome/Down syndrome are presented in table 1.

Multiple logistic regression confirmed that significantly more dual sensory impairment is diagnosed in subgroups with age 50 years and over and with Down syndrome, and showed that profound ID is also a significant risk factor (table 2). A test for trend showed a significantly increasing risk of dual sensory impairment with more severe ID ($p < 0.0001$).

Table 1 Re-weighted prevalences of dual sensory impairment (%) by age (yrs) and Down syndrome -/+

	No DS	95% CI	DS	95% CI	Total	95% CI
Age < 50	2.7	1.7 - 4.1	3.6	1.4 - 7.2	2.9	1.9 - 4.1
Age ≥ 50	7.8	5.0 - 11.5	27.8	16.5 - 41.6	11.0	7.9 -14.7
Total	4.1	3.1 - 5.7	8.8	6.1 - 14.2	5.0	3.9 - 6.2

DS *Down syndrome*
95% CI *95% confidence interval*

Table 2 Summary of multiple logistic regression model with dual sensory impairment as the dependent variable

Parameter	Odds ratio	95% Confidence interval	p-value
Degree of ID: mild*	1		
Degree of ID: moderate	1.17	0.52 - 2.66	0.702
Degree of ID: severe	2.09	0.89 - 4.92	0.091
Degree of ID: profound	4.53	1.82 - 11.26	0.001
Age < 50 years*	1		
Age ≥ 50 years	3.62	2.20 - 5.95	0.000
No Down syndrome*	1		
Down syndrome	2.37	1.42 - 3.96	0.001

ID *intellectual disability*
* *reference category*

3.2 Missed diagnoses and characteristics of group with dual sensory impairment

Dual diagnosis had been identified prior to the study in 9 out of 77 cases (12%), whereas only visual impairment had been identified in 6/77 (8%) and only hearing impairment in 24/77 (31%) cases. In 38/77 (49%), no sensory impairment had been diagnosed. One person was really functionally deaf (profound hearing impairment) and blind, whereas two persons were blind, combined with severe hearing impairment, and two were functionally deaf, combined with visual impairment. The others had less severe impairments. Six persons had severely impaired visual fields, next to low visual acuity.

The group consisted of 39 females (50.6%) and 38 males. Sixty-four (83.1%) lived in central residential settings, whereas the other 13 had been included through community-based homes or day activity centres. Age distribution is shown in Figure 1; 60 were younger than 65 years (77.9%). The level of ID was not registered in 4, mild in 8 (10.4%), moderate in 29 (37.7%), severe in 21 (27.3%) and profound in 15 persons (19.5%).

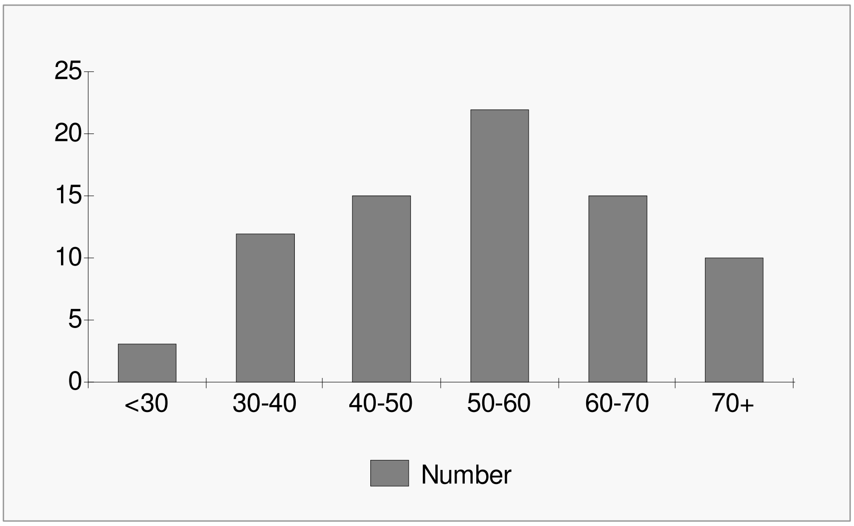

Figure 1 *Age distribution in study group with dual sensory impairment (n = 77)*

The aetiology of ID was Down syndrome in 32 cases (41.6%), whereas the following diagnoses were made in one case each: partial trisomy 2q, Angelman syndrome, CHARGE syndrome, kernicterus, perinatal brain damage, meningitis/encephalitis, mucopolysaccharidosis, and traumatic brain damage. No diagnosis of intrauterine infection had been made in this group. Etiological diagnoses were missing in 37 cases (48%).

Refraction could be reliably measured in 27 cases, slit lamp biomicroscopy was feasible in 66 and tonometry in 8 cases only. In 44 cases (57%), 24 with Down syndrome and 20 with other etiologies, treatable ophthalmological conditions were diagnosed (diagnoses in persons with Down syndrome between parentheses): severe binocular hypermetropia 1 (0), severe binocular myopia 2 (1), moderate binocular hypermetropia 5 (1), moderate monocular myopia 2 (1), advanced binocular cataract 22 (13), advanced monocular cataract 8 (2), binocular keratoconus 12 (10), monocular keratoconus 2 (1), binocular corneal opacities 9 (6), monocular corneal opacities 5 (3), ocular hypertension 2 (0). Twenty-seven had one and 17 had two or three treatable conditions. Beginning cataracts were found in 6 other persons. The type of hearing loss was sensorineural in 29 (38%), conductive in 6 (8%), mixed sensorineural and conductive in 24 (20%), and unclear in 18 cases. The audiogram curve showed a decrease towards higher frequencies in 30 cases (39%), indicating presbyacusis.

DISCUSSION

This is the first population-based study of combined visual and hearing impairment in adults with intellectual disabilities (ID), based on expert ophthalmological and audiological assessments and WHO definitions. It shows a re-weighted prevalence of 5.0% (95% CI 3.9 - 6.2%) in the total adult population, using Dutch residential or day activity ID services. The risk increases significantly with more severe ID, in the subpopulation with Down syndrome, and in case of 50 years and over. Nevertheless absolutely, 78% of identified cases were younger than 65 years. In only 12%, the complete diagnosis had been made prior to the study. In 57% treatable ophthalmologic conditions were diagnosed.

In the literature on dual sensory impairment, this condition usually is called deaf-blindness. Although this term suggests a total inability to see or hear, it is usually used for a broader range of sensory impairments, defined either functionally[19] or according to WHO definitions.[20] Therefore, we prefer the word 'dual sensory impairment'. Reported prevalences in the general population appear to be extremely scarce. They concern either children, or adults with the Usher syndrome (which is not associated with intellectual disability), or ageing people. Representative population-based studies in children on both hearing and visual impairments are few, whereas those that are available, do not report on the combination.[21,22] Prevalences of visual or hearing impairment separately appeared to be around 0.1%, so the prevalence of the combination must be (much) lower than that. Dual sensory impairment, acquired before the age of 65 years, is primarily caused by Usher syndrome, with population prevalences of 5.0 - 6.2 per 100,000 adults.[23,24,25] Based on these figures, we roughly estimate that the prevalence of dual sensory impairment in the general adult population younger than 50 years, is not higher than 0.03%. This is about 1% of the re-weighted prevalence of 2.9% (95% CI 1.9 - 4.1), found by us for the adult ID population younger than 50 years.

For age 50 years and over, we found a re-weighted prevalence of 11.0% (95% CI: 7.9 - 14.7). Recently, prevalences of dual sensory impairment in the general ageing Dutch population have been estimated from a compilation of five large national data files:[26] in the population older than 65 years, the prevalence is 1.4% (95% CI 0.8 - 2.0), and in the population older than 85 years, 4.8% (95% CI 2.4 - 7.2%). The considerably higher risk in people with ID might be partially explained by different definitions and assessment methods. Present data in Dutch national files are primarily based on self-report or proxy-report, using standardised O.E.C.D. (Organisation for Economic Co-operation and Development) questions,[27] and not on medical assessments. In one of the included data files, the Leiden 85+ study,[28] both self- or proxy-reports and clinical assessments were applied, resulting in similar prevalences. However, in that study, mild hearing impairments were not included. If in our analysis mild hearing impairments would have been excluded, too, re-weighted prevalence of dual sensory impairment in the population of 50 years and over would have been 7.4% (95% CI 4.5 - 11.2) instead of 11%. This is still impressively higher than the 1.4% prevalence in the general Dutch population aged 65 years and over. At this point however, we stress that adults with ID have less capacities than other people to cognitively compensate for sensory losses, and are more prone to unfavourable speech-noise ratios in homes and day activity centres. Therefore, for this population, we prefer to include mild hearing impairments in the prevalence figures, as a basis for health policies.

The diagnosis of dual sensory impairment was completely or partially missed in a majority. Indeed, it has been reported repeatedly, that sensory losses often are not recognised as such by ID adults, their carers and physicians.[29,30,31] Although part of congenital and childhood visual function losses, as well as age-related conditions such as macular degeneration are untreatable, treatable conditions were found in a majority. Keratoplasty is only applicable in selected patients,[32] and not every person with a severe ID accepts spectacles, but most refractive errors, cataracts and age-related glaucomas are normally treatable. The same applies to ENT surgery and rehabilitation with hearing aids. So timely detection and treatment might have relevant consequences for communication capacities, daily functioning, independence, work and social contacts of these people, and by consequence for costs of care. Is this really so? In older adults with normal cognitive abilities, communication difficulties resulting from sensory loss may lead to depression, anxiety, lethargy and social dissatisfaction,[33] whereas the combination of both hearing and visual impairment correlates to significantly lower scores of daily functioning.[34] However, no studies of disability, specifically resulting from visual and hearing impairment, and no studies of effects of treatment and rehabilitation have been published for the ID population. Different outcomes might be obtained in children and adults, or in people with mild or moderate ID and those with severe ID, supporting different policies for detection and treatment. Nevertheless, all physicians involved in medical care of people with intellectual disabilities should be aware of the very high risk of sensory impairments in these patients, and cooperate to detect and if possible treat (dual) sensory impairments.

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4 Chapter

HEARING AIDS: EXPECTATIONS AND SATISFACTION OF PEOPLE WITH AN INTELLECTUAL DISABILITY, A DESCRIPTIVE PILOT STUDY

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ABSTRACT

Background: In spite of an increased risk of hearing impairment in persons with an intellectual disability (ID), rehabilitation with hearing aids often fails. We explored which elements contribute to expectations of and satisfaction with hearing aids in adults with an ID.

Methods: Study population: 16 adults with a mild or moderate ID and a recent diagnosis of hearing impairment, Method: semi-structured interviews prior to hearing aid fitting and six months afterwards. Outcomes were related to satisfaction domains as described for the general population.

Results: Before intervention: most participants were aware of their hearing loss and familiar with reasons for hearing aids. Information on hearing aids was often remembered incorrect or incomplete. Half of them preferred invisible or, on the contrary, brightly coloured aids.

After intervention: In positive and negative experiences, all satisfaction domains as described for the general population could be recognised. Fulfilment of explicit wishes added to acceptance.

Most participants were partially or totally dependent on carers in use and maintenance of hearing aids and detection and solving of problems. A next door hearing aid supplier appeared useful.

Conclusions: Adults with a mild-moderate ID, may have explicit attitudes and wishes concerning hearing aids, and if asked, are capable of expressing these. Given information is not always understood or remembered correctly and should be checked and repeated. In satisfaction with hearing aids, the same elements play a role as in the general population which should be explicitly anticipated: benefit, cosmetics and self-image, sound quality/acoustics, comfort and ease of use, service delivery. Looks of the aids are important (invisible or brightly coloured), and may add to acceptance and satisfaction. Staff should be trained in hearing aids maintenance, and professional help (hearing aid supplier, speech and hearing therapist) should remain closely and actively involved.

Submitted

1 INTRODUCTION

In the past decades, major improvements in quality of sound and cosmetic appearance were achieved in hearing aids. Nevertheless, many persons with a hearing loss that would warrant purchase of hearing aids do not own such devices.[1,2] It appears that experienced handicap by the hearing loss especially influences acceptance of hearing aids in a positive way.[3] This experienced disability or handicap however is not directly related to the level of hearing impairment: a mild hearing loss may be experienced by some as a severe handicap and vice versa.[3,4]

Usage of hearing aids appears to be strongly related to perceived benefit.[2] On the other hand, perceived benefit is not the only issue that determines satisfaction with hearing aids. Based on a review and synthesis of published work, followed by an additional survey (structured interviews), Cox and Alexander[5] categorised the important elements in satisfaction with hearing aids in six domains: benefit, cosmetics and self-image, sound quality/acoustics, comfort and ease of use, cost, and service.

With a population-based prevalence of 30.3%, hearing impairment occurs more frequently in adults with an intellectual disability (ID),[6] as compared with a prevalence of 16 - 17% in general adult populations.[7,8,9] However, rehabilitation with hearing aids often fails in these people. It is often assumed that people with an ID do not easily accept hearing aids. We therefore wanted to explore in this group which elements contribute to expectations of and attitude towards hearing aids, and to satisfaction after the fitting. This in order to find out whether these elements are comparable with those in the general population, or are specific for persons with an ID.

2 POPULATION AND METHODS

2.1 Population

Sixteen adults with a mild (IQ 55 - 70) or moderate ID (IQ 35 - 55), in whom hearing impairment had been recently diagnosed, were recruited from an implementation study of audiological rehabilitation in ID service-providers, which was in progress in 2002. Written informed consent was obtained from the participants and/ or their legal representatives.

Hearing impairment was defined as a loss of more than 25 dB at the best ear, averaged over three frequencies (1, 2 and 4 kHz). The degree of hearing impairment was classified as mild (losses of 26 - 40 dB), moderate (41 - 60 dB), severe (61 - 80 dB), or profound (over 80 dB).[10] Indication for hearing aids: Pure Tone Average over 25 dB.

2.2 Methods

The rehabilitation programme has been reported elsewhere, and included information booklets,[11] hearing aid prescription, habituation and training, staff training and assessment of acoustic conditions.[12] All participants were interviewed individually by the researcher before, and six months after the introduction of hearing aids. The topics of the semi-structured interviews were related to the research questions (Appendix A). Apart from an open interview technique, the interviewer posed short simple questions, and asked the clients to illustrate their answers whenever this was relevant. During the interview, information was regularly checked with the interviewee if it was correctly understood. In case of answers regarding colours, it was checked if the participant could correctly point out the specific colour. Each client was tested on acquiescent responding (Appendix B), which means the tendency of providing the interviewer with the answers assumed to be most desirable. The interviews lasted around 10 minutes each, and were recorded on tape after consent of the interviewee. The taped interviews were transcribed literally by an independent co-worker. Results of the interviews were described per research question. Positive and negative experiences were compared with the earlier mentioned domains of satisfaction[5] which have been developed for the general adult population: benefit, cosmetics and self-image, sound quality/acoustics, comfort and ease of use, cost, and service.

3 RESULTS

3.1 Population

Of the sixteen clients who participated in the study (table 1), two (J and L) were not able to give relevant answers. Hearing aids turned out to be a success in J, and a failure in L. Because of this, results are based on the interviews of 14 clients, six females and eight males in the age range of 30 to 67 years. Nine had a mild, and five a moderate hearing impairment. Nine participants lived in community-based homes, and five in larger residential settings. Two of them (I and K) were not interviewed prior to the hearing aid fitting, because of psychological and logistical reasons. Prior to hearing aid fitting, two participants with a moderate ID gave relevant answers to a few questions only; this improved noticeably during the second interview after the hearing aids had been introduced (table 2). Acquiescent responding could be established in none of the 14 participants.

Acoustical conditions were unacceptable, but not timely improved, in the living rooms of participants C, F, G, H, and K, and not assessed in living rooms of participants M, N, O, and P. A speech and language therapist to provide hearing aid habituation and training was not available for participants K, M, N, O and P.

Table 1 Characteristics of the participants, their hearing function and hearing aids

	sex	age	interval start HA- interview	type of care*	level of ID	H.I. in dB right	H.I. in dB left	class. H.I.	hearing aid	Nr. of H.A.	S & L ther.
A	♀	48	6 months	inst	moderate	38	43	mild	analogue	2	+
B	♂	54	6 months	inst	moderate	75	42	moderate (R > L)	analogue	1 on L	+
C	♂	45	6 months	inst/cb	mild - moderate	27	40	mild	analogue	1 on L	+
D	♀	36	6 months	inst	moderate	27	28	mild	analogue	1 on L	+
E	♂	60	6 months	inst	moderate	35	82	mild (L > R)	analogue	1 on R	+
F	♂	54	6 months	inst/cb	mild	38	42	mild	in-the-ear	2	+
G	♂	55	6 months	inst/cb	moderate	38	37	mild	digitital	2	+ → -
H	♀	54	6 months	inst/cb	mild - moderate	37	48	mild	digitital	2	+
I	♀	43	4 months	inst	moderate	60	55	moderate	hybride	2	+
K	♂	30	6 months	inst/cb	mild	38	48	mild	hybride	2	-
M	♀	47	1 month	cb	mild	50	50	moderate	digitital	2	-
N	♀	66	6 months	cb	moderate	62	47	moderate	digitital	2	-
O	♂	67	6 months	cb	moderate	52	58	moderate	digital	2	-
P	♂	38	6 months	cb	mild	37	43	mild	digital	2	-

Excluded participants because no relevant answers were given:

J	♂	57		inst	moderate - severe	43	50	moderate	hybride	2	
L	♀	60		cb	moderate	45	45	moderate	digital	2	

inst	*institutional based*
cb	*community based*
inst/cb	*living in the community but using the facilities of the nearby institution*
class.	*classification*
H.I.	*hearing impairment*
H.A.	*number of hearing aids*
S & L ther.	*Speech and Language Therapist*

3.2 Characteristics of hearing aids prescribed

Ten participants were prescribed binaural, and four unilateral hearing aids (B, C, D and E) because of asymmetrical hearing loss or insufficient hearing function in the other ear.

Six persons received analogue, six digital, and two hybrid hearing aids (table 1). All clients were primarily prescribed 'behind the ear' hearing aids. Because of his persistent attitude, client F received 'in the ear' hearing aids. He insisted on having these because of his glasses.

3.3 Results of the interviews before fitting with hearing aids (n = 12)

In table 2 is shown for each topic which participants gave (relevant) answers.

Table 2 Participants who gave relevant answers (*)

		A	**B**	**C**	**D**	**E**	**F**	**G**	**H**	**I**	**K**	**M**	**N**	**O**	**P**
3.3.1.1 awareness															
3.3.1.2 booklet	not familiar with the booklet			+	+				?	ni	ni	?	+		+
3.3.1.2 why HA		+	+	+	+	+	+	+	+	ni	ni	+	+	+	+
3.3.1.2.number of HA	did not know	+	+	+	+		+	+	+	ni	ni	+			+
3.3.1.2 looks of the HA	did not know			+			+	+	+	ni	ni	+	+		
3.3.1.2 how HA help	did not know		+	+	+		+			ni	ni	+			
3.3.1.3 attitude	no relevant answer	+		+	+		+	+		ni	ni	+	+		+
3.3.1.4 colour preference	did not know	+	+	+	+	+	+	+	+	ni	ni	+	+		
introduction of hearing aids															
3.3.1.2 booklet	not familiar with the booklet	+	+	+	+				+	+	+	+	+		+
3.4.1.1 benefit		+	+	+	+	+	+	+	+	+	+	+	+	+	+
3.4.1.2 looks of the HA		+	+	+	+	+	+	+	+	+	+	+	+	+	+
3.4.1.3 environmental sound		+	+	+	+	+	+	+	+	+	+	+	+	+	+
3.4.1.4 comfort		+	+	+	+	+	+		+	+	+	+	+	+	+
3.4.2 dependence on carers		+	+	+	+	+	+	+	+	+	+	+	+	+	+

The numbers correspond with the paragraphs,

ni	*not interviewed prior to the hearing aids*
?	*unclear if booklets were familiar*
HA	*hearing aids*

3.3.1 *Which elements contribute to expectations and attitude towards hearing aids?*

3.3.1.1 Awareness of hearing loss. Ten interviewees were aware of their hearing loss. A denied it, and M was unclear about it . She indicated however her preference for sitting at the 'table for deaf people', because "that is with gestures, and there I can express myself better".

3.3.1.2 Information on hearing aids. Prior to the fitting of the hearing aids 4 out of 12 participants were familiar with the information booklet. After the first interview, booklets were again handed to the carers of the participants. This resulted in 10 out of 14 participants being familiar with the booklets during the second interview. Although one of them thought the booklet to be worthless and childish (C), and another found it difficult, the other eight said they appreciated it, which was expressed as: nice, good, or interesting. Three of them could illustrate this: due to the booklet they understood things better (N and P) and it gave practical information (H).

All 12 clients knew why they would receive hearing aids. It was either 'my ear doesn't work', 'not hearing well' or 'to hear better'. The number of hearing aids was mentioned correctly by five, incorrect by four, and was unknown in three persons. Six knew what hearing aids looked like (based on experience with hearing aids in parents or group members). One participant (E) assumed that the hearing aid would be put under the skin (he pointed at his chest) and from there linked to the ear with a wire. Five clients knew what hearing aids would do for them. It was either that the sound would be louder (C and M), or the expectation to hear better (B, D and F). One of them (D) added to this: "A little wire that will move your ear". F had heard he would have the hearing aids on trial first and was worried that he would be used as a guinea pig.

As a conclusion, all participants knew why they would receive hearing aids, but information on the looks, possible benefit and number of hearing aids was remembered incomplete, incorrect, or twisted in the majority of participants.

3.3.1.3 Attitude towards hearing aids. Answers to this question are shown in table 3. Three participants were (moderately) positive (F, G and P), whereas three others had a more negative response which had to do with comfort (C) and a fear of the lifelong aspect of the fitting (D), whereas client A could not specify her attitude any further. For client M invisibility was important.

3.3.1.4 Colour preferences in hearing aids. Four participants preferred brightly coloured hearing aids (blue and red), one a grey one, two were satisfied with brown, two had no preference and two did not know. One client could not correctly point out the right colour.

Table 3 Attitude towards hearing aids before the start with these devices

A	"Don't like it that I will get hearing aids" (could not specify this further).
C	"If they trouble me, I won't wear them, I won't wear them".
D	"I am afraid that I will have to live with them all my life".
F	"I like it, because I know that when I will have a little thing like that, I will hear somewhat better".
G	"Give it a try; old age I think".
M	"I want that it cannot be seen, but that there is one in it alright".
N	"On the 11th I will have to go to the ear doctor and then they can check if I need a hearing aid, or maybe not".
P	"Honestly I am glad that I will receive it, because then I will be able to hear a lot better".

3.4 The interviews after hearing aids were received

3.4.1 What are positive and negative experiences with hearing aids?

3.4.1.1 Benefit of the hearing aids. In table 4 reasons are described that contributed to success and failure. Hearing aids were a success in 11/14 clients (3 mild ID, 1 mild-moderate ID, 7 moderate ID). With the exception of one, all these clients received hearing aids on both sides. As reasons for success were mentioned: hearing (other persons) better (A, F, H, I and O), understanding other persons better (D, K and N), understanding better what is said on television and radio (B) and improved detection of specific sounds (F: oncoming traffic and the cat). In addition to understanding other persons better, client P described the decrease in fatigue which enabled him to work whole days instead of half.

A special case is G (table 3) in whom hearing aids were an initial success after they had been introduced under the guidance of a speech and language therapist. During a consultation, the audiologist had increased the volume of the hearing aids, which was too loud for G. The hearing aids had disappeared in the drawer; until the moment of the interview (2½ months later), no action had taken place to solve this problem.

Hearing aids fitting failed in three participants (C, E, M). E and M did not experience any benefit, whereas in C, irritation of the device in his ear and whistling dominated his experience; no benefit was reported or denied.

3.4.1.2 Looks and colour of the hearing aids. Twelve participants were content with the looks of the hearing aids, which was expressed as 'nice' or 'good', or satisfaction with the small size. E and M, in whom hearing aids failed, disliked the visibility of the hearing aids. All participants received skin-coloured hearing aids. Ten were content with this colour, although three of them preferred red ones prior to the fitting. Two (B and O) now preferred brightly coloured ones (blue and red) and two (I and M) had no colour preference.

Conclusion: cosmetic preference is not only aimed at invisibility, but also at beautification of the hearing aids.

3.4.1.3 Environmental sound. Eleven participants experienced problems with environmental sounds (table 5). Eight were troubled by the loudness of sounds, caused by other people's voices (A, D, K and P), by the television-set or radio (F and M), or by everything (H and G), which resulted in a failure in G (see also § 3.4.1.1). Two of them were also troubled by persons talking at the same time: D did not understand what was said, and K found it very tiring. B was troubled by the sound of the wind. Three (A, C and N) were disturbed by humming or whistling sounds when other people were talking or when the television was switched on. Three participants experienced no trouble with sound (E, I and O).

As a conclusion, eleven participants experienced problems with environmental sounds which in the majority had to do with loudness. In five of these cases, acoustics in the living rooms had been diagnosed as unacceptable.

Table 4 Reasons for success or failure of hearing aid fitting

	Success or failure	**reason success or failure**
A	+	"I can hear better now. Sound of people. If they talk I can understand them".
B	+	"I can hear better what they say, on television and on the radio. If the hearing aid is out of my ear, then I cannot hear so loud, very soft. I want a hearing aid just like this for the other ear".
C	-	"The hearing aid wasn't comfortable, it just irritated me. So I said: I take it off and will not put it in for the time being".
D	+	"I can understand staff better, that's important".
E	-	He could not tell the difference with or without. "I think it is a waste of money." Apart from this he thought the hearing aid to be too big. "People were staring at me".
F	+	"I think they are perfect. I had asked for these ('in the ear' hearing aids) you know". One hears better, I can hear the oncoming traffic better. And if my cat is miaowing a few houses down the block, then I know exactly: that's my cat".
G	+ → -	Hearing aids did help him at the start but: "Some bloke* messed with them. I am not going to mess with your (interviewer) things either, put things loud. It drives me crazy".
H	+	"I can hear better".
I	+	"Without hearing aids I can not hear it, hearing aids make you hear other people better".
K	+	"Can hear better what we are talking about".
M	-	She found it a nuisance when hearing aids were inserted in the ears and were taken out again. Apart from this she thought she would receive one hearing aid only. "They are in an envelop in the hearing aid shop, but I won't wear them, I'm not going there anymore for these aids, if I can communicate well like now" "It was louder with the hearing aids. But I can talk more easily without them" "I didn't like these things behind my ears. I want to have something that you cannot see. I would like to give that a try".
N	+	"Carers had to say things a few times, and I said: I don't understand it. Now I understand everything".
O	+	"If they are in my ear, I can hear more easily, if somebody starts to talk to you I can hear it more easily".
P	+	"If somebody says something to me, I can hear things better". "I work on a school as a caretaker. Before, I had to copy something, say two or three times, and then I came back with five or six. And then they said: no, we've ordered two or three. And then I said: I didn't hear it" From January I worked half days, April 1st I received my hearing aids, …. and since fourteen days I am back to working all day".

* See § 3.4.1.1

Table 5 Hearing aids and environmental sounds

Participant	
A	"When you are talking to somebody, then it's really loud and it hums". "If the radio is turned on, if there's music, it hums".
B	He is troubled by the sound of the wind when he is wearing his aid: "When I have to do some shopping (on his bicycle), then the wind goes hoeoeoe, but the wind doesn't bite".
C	It started to whistle and to hum in the living room when he was with others. Then he removed it from his ears.
D	"If I go to my work and put it in, then I hear people talk very loud. I rather have soft language. Sometimes I hear my neighbour talk very loud, then I have these fears, but then I just look for staff" "If people are talking with each other, I can't hear anything".
E	No trouble with sound.
F	They make a lot of noise in the bus; however in a touring bus they do not. He explains that the engine in a touring bus is at the back instead of in the middle, as in an ordinary bus. He does not wear the hearing aids in the kitchen where he works because of the noise. He is troubled by loud music in the living room and then asks if the volume can be turned down.
G	see § 3.4.1.1
H	"If I wear them, everything sounds so loud". "At my work during coffee break and with sowing and hammering I turn them off. ". If there is a little noise, like (coffee)cups and so on, then I take them out of my ears".
I	No trouble with sound.
K	"At the fair it usually hurts a lot, because of the very big music boxes" "When people are talking at the same time, it is very tiring". "I like to simply listen at people, but they shouldn't have loud voices. Then I say: take it easy".
M	She does not like thunderstorm and ambulances with wailing sirens or television sounds that are too loud.
N	"When the television is too loud I hear cracking, cracking noises".
O	No trouble with sound.
P	"If somebody talks loudly, they are used to do that of course, then I say: not too loud".

3.4.1.4 Comfort. Nine participants experienced trouble with the hearing aids, which had to do with pain (B, N and P), discomfort (C and M, see also § 3.4.1.1), and humming or whistling sounds (A, C, E, I and P) which persisted in four of these five after the hearing aid was inserted in the ear. Participant D removed the hearing aids during meals because of fear that the ears would start itching afterwards. On participant G's trouble is already reported in § 3.4.1.1. Four participants (F, H, K and O) experienced no discomfort.

Two of the three participants with complaints of pain, and one with persistent whistling attended their hearing aid supplier, whose shop was next door, and who successfully "did something about it". In the third participant with pain, the problem still existed at the time of the interview, 6 months after the hearing aid fitting.

As a conclusion, 9/14 participants experienced problems related to the comfort of hearing aids. Action was taken in three participants only. These participants belonged to a group of four who lived next door to a hearing aid supplier.

3.4.2 *To what extent do participants depend on carers?*

In table 6 participation of the participants in the use and maintenance of hearing aids is described in relation to the degree of ID. Six clients were totally dependent on carers, six partly, and only two were totally independent.

Table 6 Participation of clients in the use of hearing aids

Participation in use and maintenance	Mild ID	Moderate ID	Mild-moderate ID
None	1	4	1
Taking off HA		1	1
Putting in and taking off HA		3	
Everything except connecting hearing aid and ear mould after cleaning	1		
Independent of carers	2		

HA *hearing aids*
ID *intellectual disability*

4 DISCUSSION

This pilot study shows that persons with a mild or moderate intellectual disability (ID) are capable of expressing their opinions on, and experiences with hearing aids, provided that simple questions are asked on concrete matters.

Most participants were aware of their hearing loss and familiar with the reason for hearing aids prescription. Expectations with regards to the looks and possible benefit were based on information that was remembered incomplete or incorrect in a majority of participants; this in spite of information booklets that were distributed through staff. Information before and during hearing aid fitting should be repeated and checked to improve its effectiveness.

It is remarkable that two out of three participants in whom hearing aids failed, had expressed explicit wishes with regards to the hearing aids concerning comfort and cosmetics, which apparently could not be met; these wishes were mentioned as reasons for failure afterwards (table 4). On the other hand, in the one participant who was able to persist in his demand towards in-the-ear hearing aids, the fulfilment of this has certainly contributed to his satisfaction. This stresses the importance of exploring clients' attitude prior to the fitting, including explicit wishes about the hearing aids.

This study also shows that elements that are central to satisfaction with hearing aids in the general population (benefit, cosmetics and self-image, sound quality/ acoustics, comfort and ease of use, and service)[5] can also be recognised in the positive and negative experiences of this study group. Reported benefit from the hearing aids had to do with improved hearing, improved understanding of other persons, or improved hearing of radio or television, but also with improved detection and localisation of other sounds. These items are comparable with the five factors that Kramer et al[13] described as fundamental in auditory disability: distinction of sounds, auditory localisation, intelligibility in quiet as well as in noise and detection of sounds. In addition, the fatigue that markedly diminished in one participant (table 4), is also described by Kramer et al[14] and by others[15,16] as adding to disability, because of the constant effort to hear and respond appropriately.

Cosmetic aspects did not only appear in an explicit wish for invisibility, but in some participants in the opposite desire for brightly coloured devices. Giving clients a choice might positively influence acceptation of the device. We do realise that 'in the ear' hearing aids may not be an option to every person with an ID, because of the more complicated handling and vulnerability, but may be an option in selected clients, contributing to satisfaction and acceptance.

'Quality of sound and acoustics' also appeared to be an important domain which predominately had to do with loudness (8/12). The poor acoustical conditions in the houses of four of these eight participants will certainly have contributed to the problem. This aspect is also included in the adversiveness score in the Abbreviated Profile of Hearing Aid Benefit[17] and described by Kramer et al[13] as intolerance of noise. More attention should be given to the described complaint which can be solved by adjusting the Wide Dynamic Range Compression in the hearing aids and by environmental noise reduction.

In this domain as well as in the domain 'comfort and ease of use', dependence on carers became apparent. Six months after the hearing aid fitting, problems concerning loudness of sounds, and discomfort and pain still existed in eight and six participants respectively, in spite of the training of staff. For the detection and solving of these problems but also for the daily use and maintenance, even persons with a mild ID appeared to be dependent on carers. However, community-based ID services in the Netherlands do not always provide specialised healthcare for their clients. Carers have a social-educational background, whereas not in all ID services specifically experienced speech and language therapists are available to provide hearing aid habituation and training.[12] That the hearing aid supplier lived next door to three of the studied participants who had their problems solved, will certainly have facilitated their initiative to take action.

CONCLUSION

Adults with a mild-moderate ID may have explicit attitudes and wishes about hearing aids, and if asked, are capable of expressing these. Given information is not always understood or remembered correctly and should be checked and repeated. In satisfaction with hearing aids, the same elements play a role as in the general population which should be explicitly anticipated: benefit, cosmetics and self-image, sound quality/acoustics, comfort and ease of use, service delivery. Looks of the aids are important (invisible or brightly coloured), and may add to acceptance and satisfaction. Staff should be trained in hearing aids maintenance, and professional help (hearing aid supplier, speech and hearing therapist) should remain closely involved.

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Appendix A Topics of the interview before and after the start with hearing aids (translated from Dutch)

Interview before the start with hearing aids

1. I am told that you will receive hearing aids. Whý will you receive hearing aids?
2. Did you notice yourself that you can't hear properly?
3. How will those hearing aids help you?
4. What do hearing aids look like?
5. Would you like to receive coloured hearing aids?
6. Will you have one or two hearing aids?
7. What do you think about the hearing aids that you will receive?
8. Who told you that you will receive hearing aids? What did he/ she tell you?
9. Did your carer show you a booklet on hearing aids? Can you remember what is in the booklet? Can you describe a picture in the booklet? What do you think of the booklet?
10. Do you know other people with hearing aids?
11. What do you think about those hearing aids
12. Do you have any questions about the hearing aids?
13. Do you have anything to add about the hearing aids?

Interview after the introduction of hearing aids

1. What do you think of your hearing aids?
2. Do the hearing aids help you?
3. Do you wear the hearing aids all day?
4. What do you think of the way the hearing aids look like?
5. What do you think of the colour of the hearing aids?
6. Are you familiar with this booklet (booklet is shown)? Can you remember what is in the booklet? Can you describe a picture in the booklet? Did the booklet help you with your hearing aids?
7. What do other people think of your hearing aids?
8. How does it feel to have the hearing aids in your ears?
9. Do the hearing aids hurt you every now and then?
10. Are there sounds that bother you?
11. Who puts the hearing aids in your ears?
12. Who takes the hearing aids out of your ears?

Appendix B Client testing for acquiescent responding[18] (Cummins 1997)

Procedure

The primary carer may be present for the administration of the test. However, if they are present it is essential that they:

1. Be instructed to make no response whatsoever to the questions as they are read to the client.
2. They must be located outside the client's visual field.

Administration

After checking that the carer is informed as above, and the client is comfortable and ready to respond, carefully and slowly read each question:

1. Point to the client's watch or some item of clothing.
   "Does that (watch) belong to you?"
2. Do you make all your own clothes and shoes?"
3. Have you seen the people who live next door?"
4. Did you choose who lives next door?

Scoring

If a positive response is provided to items 2 and 4, no further testing should take place.

5 Chapter

IMPROVING THE QUALITY OF AUDIOLOGICAL REHABILITATION IN SERVICES FOR ADULTS WITH INTELLECTUAL DISABILITIES

PART I

DESCRIPTION OF AN IMPLEMENTATION

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ABSTRACT

Background: People with an intellectual disability (ID) have an increased risk of hearing impairment, but audiological rehabilitation is often lacking or failing. No systematic research has been published on audiological rehabilitation of people with an ID.

Study objective: to study the feasibility of the implementation of an audiological rehabilitation protocol in ID centres, and to describe the practical barriers and facilitating initiatives that are met during the implementation process.

Methods: A multidisciplinary protocol for audiological rehabilitation in people with an ID was developed. Adults with a recent diagnosis of hearing impairment and an indication for treatment were recruited from five ID services. Project requirements (local project coordinator, organisational and financial investments) were discussed with the management prior to participation. After identification of factors that might facilitate or hamper the implementation process, comprehensive implementation strategies were developed. In the implementation process a preparatory phase, an intervention phase, and an adoption phase were distinguished. Barriers and facilitating initiatives were identified in relation to management, the professionals, the social context and the organisational context.

Results: In only 3 out of 31 included participants criteria were realised for completion of the intervention phase. Barriers were identified in relation to management (prolonged decision procedures to participate, other priorities for acoustical adjustments and trainer, temporary assignment trainers, insufficient time for speech and hearing therapists, lack of time for, and insufficient expertise in project management), in relation to the audiologists (assessment of acoustical conditions no task for audiologists, not equipped enough to assess acoustical conditions), in relation to the social context (poor cooperation between disciplines), in relation to the organisational context (absence of speech and hearing therapist, change of house, no budget for acoustical assessment), and in relation to the local project management (communication problems). Facilitating initiatives were encountered in the professionals in the form of initiatives to ensure continuity.

Conclusions: Implementation of an audiological rehabilitation programme in ID centres turned out to be not feasible within a period of 2;3 years. The implementation process was seriously hampered by severe delays in audiometry. Barriers were already met at an organisational level of audiological and ID services instead of at the expected client-carer level. Initiatives to ensure continuity of parts of the protocol were taken by professionals. The required expertise of and time for local project management was severely underestimated by management.

A concise report of chapter 5 and 6, titled 'Audiological rehabilitation in adults with intellectual disability: why does it fail?', has been published in the Journal of Policy and Practice in Intellectual Disabilities 2005; 2:66-67.

1 INTRODUCTION

Although children and adults with an intellectual disability (ID) are at an increased risk of hearing impairment, this additional impairment remains unrecognised and untreated in many cases.[1,2,3,4,5,6]

Screening programmes were started in the Netherlands after the publication of Dutch diagnostic guidelines for this group.[7] This has resulted in the detection of many cases of previously unidentified hearing impairment in children and adults.

At present, when hearing impairment is diagnosed in people with an ID, treatment and advice is given by E.N.T. specialists and audiological centres in a similar way as for people with normal intelligence. In practice the treatment of hearing impairment for people with ID quite often fails or does not even get started with. The focus of this paper is to look into why this happens, and specifically into the organisational aspects.

The poor pickup of rehabilitation programs may be due to client related factors or affected by the persons in their surroundings (care-givers). No research has been done to investigate barriers for treatment in this specific group. Therefore a prospective, descriptive implementation study within Dutch ID services was set up, aiming at adults with a recent diagnosis of hearing impairment.

The following research questions were formulated:

1. Which practical barriers and which facilitating initiatives are met in the implementation process?
2. Is adequate audiological rehabilitation in ID centres feasible?

2 SUBJECTS AND METHODS

2.1 Population

Our study aimed to implement optimal audiological rehabilitation in 100 adults with mild to severe intellectual disabilities, with an equal distribution of participants from residential care and community based homes. The participants were recruited from ID services where during the year 2000 screening of the hearing function took place. Those who failed the screen were given a referral for the district audiological centre; general practitioners or ID physicians were usually responsible for the referral. Only patients were included from those ID services where audiological diagnostic was expected to be completed during the year 2000, and where identification of hearing impairment was expected in at least five clients.

All adults were recruited with a mild to severe ID who were identified with a hearing impairment defined as a loss of at least 35 dB in the best ear, measured at 1, 2 and 4 kHz. In cases of hearing impairment between 25 - 35 dB in the best ear, a recommendation for intervention and subsequent inclusion in the study depended on the person's communication skills and needs.

Originally we also intended to develop instruments to measure effects of treatment in this population. But it turned out that we could not include a sufficient number of participants for this part of the study. Because of the original aims, the following exclusion criteria were formulated for individual participants: autism (diagnosed in three steps by means of AVZ-R,[8] AUTI-R,[9] and if indicated, clinical diagnosis), visual acuity below 0.3, and the use of a hearing aid in the past during 6 months or longer.

2.2 Study design

In a prospective, descriptive 3-year study design, the whole first year was reserved for preparations. Whereas a four-module rehabilitation protocol had already been developed before the study period, we now developed guidelines for acoustics and a training course for participating audiologists (more details in paragraph 2.3). Further, a training course for staff was designed, using a train-the-trainers model, as well as a training course for speech and hearing therapists, and a course for behavioural scientists and physicians from the ID service (more details in paragraph 2.3). After identification of factors that might hamper or facilitate the implementation process, comprehensive strategies were developed for the implementation in the ID centres (more details in paragraph 2.4).
The design included three phases:

1. In the preparatory phase (the first year), audiometry and hearing aid prescriptions were completed in participating ID services, after which inclusion of participants with hearing impairment was started. Interventions were prepared.
2. In the intervention phase (first three months of the second year) all interventions, necessary for a successful start of the fitting with hearing aids had to be completed: training courses, improvement of the acoustical conditions where necessary, discussion of communicative advices, and training of carers. This phase ended with the introduction of the hearing aids in combination with the hearing aid habituation and auditory training.
3. The adoption phase: the rehabilitation programme was considered to be successfully implemented when it would be adopted by the professionals involved, which would be shown by a permanent change in their behaviour, and structural embedding in the organisation ensuring continuity. This was investigated by means of a 9-month follow-up of the hearing impaired participants in the adoption phase, including unexpected check-up of the hearing aids and reports on the hearing aid fitting, and further by means of interviews and questionnaires in all disciplines involved.

During the preparation and intervention phase, any barrier was registered by the researcher, detailed minutes of every meeting in the ID services or audiological centres were made, and notes of every relevant contact in person, by telephone or by E-mail kept. Barriers and facilitating initiatives were identified in a descriptive way and related to management, to the professionals, to the social context, and to the organisational context.

2.3 The audiological rehabilitation protocol

In preparation of the study in 1999 a Dutch multidisciplinary team discussed barriers in the audiological care of people with an ID, based on their practical experience. The team was chaired by Verschuure and Evenhuis, and consisted of representatives from audiological centres, specialised centres for the deaf and hearing-impaired, and ID centres. Table 1 shows barriers identified, and adjustments of normal audiological rehabilitation procedures, considered necessary by the team.

Table 1 Barriers in audiological care for persons with an intellectual disability and necessary adjustments

Barriers		Adjustments needed
Lack of data on hearing impairment	→	Hearing screening
Audiometrical data of poor quality due to problems in co-operation	→	If necessary, use of objective audiometry
Too much reverberation and background noise in living rooms and day-care centres	→	Optimisation of acoustical conditions
Habituation to hearing aids may take a longer time	→	Hearing aid habituation and auditory training by an experienced speech and language therapist
Augmentative communication may be needed	→	Communicative intervention
Carers have no knowledge of hearing impairment and use and maintenance of hearing aids	→	Training of carers

Dutch guidelines for early identification of hearing impairment already had been published in 1996.[7] Based on the identified barriers, a rehabilitation protocol was developed which would be implemented in this study.

The protocol consisted of four modules:

1. Fitting with hearing aids, hearing aid habituation and auditory training
2. Optimisation of acoustical conditions
3. Communicative intervention
4. Training of carers

Figure 1 shows the four modules and the tasks which had to be initiated externally (by audiologists and researcher) above the line, and subsequently the tasks the tasks that had to be performed by staff from the ID services below the line.

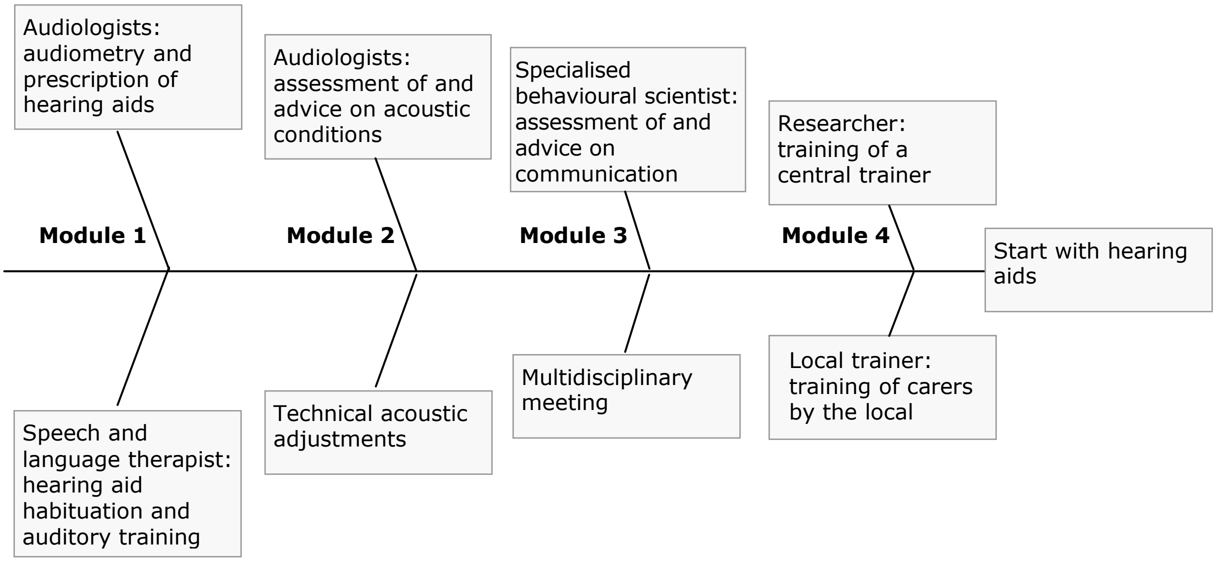

Figure 1 *The audiological rehabilitation process*

Ad Module 1 *Hearing aid fitting, hearing aid habituation and auditory training*
Innovative in this module is that the audiological service offered audiometry as well as fitting of the hearing aids on site of the ID centres instead of in the audiological centres, at least to those with a more severe degree of ID. For clients with a mild ID exceptions could be made when these people would be able to follow the usual lines. Hearing aids, prescribed by audiologists or ENT specialists, were provided by hearing aid suppliers. The diagnostic and rehabilitation protocols were discussed at the level of the Federation of Dutch Audiological Centres (FENAC, a cooperative umbrella organisation for the Dutch audiological centres) and accepted.

Another innovative element in this module is the hearing aid habituation and auditory training. In this study it would be provided by the speech and hearing therapists in the ID services in cooperation with the audiological centres. Additional training was provided in fitting with hearing aids and hearing aid habituation in order to obtain the necessary expertise (see also table 2).

Criterion for completion of the intervention phase of module 1: Hearing aids prescribed, and hearing aid habituation and auditory training available by a speech and hearing therapist who followed the training course.

Ad Module 2 *Optimisation of acoustical conditions*
A problem that so far had not been explicitly identified, was reverberation in the living rooms of community-based homes and in day-care centres for people with ID. This is caused by the size of the rooms, which in case of group-based living conditions may be larger than usual, and by the choice of interior decoration materials, which are often acoustically reflective because of hygienic demands and wheelchair use (hard floors and few soft furnishings). Reverberation causes verbal communications to become a background noise. Poor speech intelligibility in a noisy environment is one of the most common problems for people with sensorineural hearing impairment.[10,11] These studies show that people with sensorineural hearing

loss require a better signal-to-noise ratio for understanding speech. If there is too much reverberation in the room, the reverberation itself can be considered a background noise and this background noise makes speech from a normal distance unintelligible for people with a hearing loss. Hearing aids amplify sounds and make them audible. Amplification does not change the signal-to-noise ratio greatly, making speech and the noises very audible without improving the intelligibility of the speech. Provision of hearing aids to people living and working in strong reverberant conditions therefore is useless. It is therefore essential that unfavourable acoustical conditions are adjusted, as this in itself may cause a failure of the audiological rehabilitation. However, first of all guidelines had to be developed for acoustics in group homes and day-care centres. This was performed by Verschuure (Dept ENT/ Audiology, Erasmus Medical Center Rotterdam) in collaboration with Nijs and Van Berlo (Faculty of Architecture, Technical University Delft), based on existing knowledge on the required threshold signal-to-noise ratios for hearing-impaired people[12] and on the relationship between speech intelligibility and room acoustics. A workshop and training course on the assessment of and advice on acoustics were then organised for audiologists, involved in the study. The workshop presentations were published (in Dutch), together with a computer diskette containing an inventory of characteristics and costs of different sound-absorbing materials, available in the Netherlands.[13]

Criterion for completion of the intervention phase of module 2: Speech Transmission Index values (a measure for speech intelligibility) in the living rooms of at least 0.7.[13]

Ad Module 3 *Communicative intervention*

As part of the rehabilitation protocol, behavioural assessment of communicative skills and needs was performed. Based on this, advices were given towards the hearing aid habituation training, and on the necessity of augmentative or alternative communication methods. During the study, this innovative element in the rehabilitation protocol was offered by behavioural scientists from specialised organisations for deaf and hearing impaired. Results of the assessment and subsequent advice were discussed during a multidisciplinary meeting in the ID service.

Criterion for completion of the intervention phase of module 3: Behavioural advice on communication given and discussed during a multidisciplinary meeting.

Ad Module 4 *Training of carers*

In order to provide optimal care, staff needs to know the consequences of hearing impairment on communication and behaviour, and how to deal with this. In addition they have to become aware of the benefits and limitations of hearing aids. And last but not least, they need the expertise for the use and maintenance of hearing aids, including troubleshooting, because many people with an ID depend on them for this. This means that carers need to be trained in order to obtain this expertise.

Criterion for completion of the intervention phase of module 4: Completed training for at least half of the carers of a team involved with an individual participant.

2.4 Strategies for implementation

With advice from an expert in quality care and a senior manager of a non-participating ID service, we developed a plan for implementing the four rehabilitation modules in the participating ID services. To this end, seven 'disciplines' were distinguished, that would be involved in the implementation: management, physicians, speech and language therapists, behavioural scientists, carers, family of participating clients, and the participants themselves. Objectives for the implementation were specifically formulated for each discipline e.g. for carers: expertise in hearing impairment and its treatment. Then, anticipated barriers were defined per discipline, based on the clinical experiences of the research group. The current situation was analysed per discipline, using a SWOT analysis (strengths, weaknesses, opportunities, threats). Subsequently, strategies based on educational, marketing, behavioural, and social interaction approaches[15] were chosen for each discipline (table 2).

2.5 Details of our approach

A practical scenario for the implementation process was developed:

a. Senior management would receive detailed personal information about the implementation project, its value for the quality of care, and its organisational and financial consequences, prior to the decision to participate. Technical acoustical adjustments, and extra hours for staff to be trained on-site had to be financed by the ID service-providers themselves.
b. Local project coordinators were identified, they would coordinate the implementation process within the ID centres, supported by the researcher (helpdesk).
c. Agreement on responsibilities and tasks according to the scheme shown in figure 1.
d. Development of competence by training
e. Assessment of and advice on communication skills and needs were performed by specialised behavioural scientists from services for the deaf and hearing impaired.

Table 2 Examples of expected barriers for implementation and strategies for improvement

Party	**Barrier**	**Strategy**
Management	Lack of time to read information material	Information material short but to the point
	Due to changes in the organisation of care, management is insufficiently motivated and has other priorities	Raise interest: - description of successful cases of treatment of hearing impairment in people with ID Emphasize advantages of the project for the care organisation, e.g. - improvement of quality of care - innovative character - diminishing of behavioural problems in clients
	Cost	Organise pressure: - involve influential people within the ID centres, e.g. physicians - publicity through family associations
Carers	No knowledge of hearing impairment and its treatment	Education and training course
	No interest in theoretical information	Inter-active, experience-aimed training by a respected colleague-carer
	Training and information not assimilated in one course	Training in two parts with observational tasks in between Additional training on the work floor Attractive, compact instruction material including sections on how to solve problems Reminder systems, e.g. sticker above the wash basin on handling of hearing aids
	No insight into effects of treatment for client and carers	Emphasize expected effects during training in a realistic perspective
	High turn-over of carers	'Train the trainers' model: so expertise in training remains available within the organisation Repetitive training Researcher to function as a help-desk during the study
	Vacancies	Not solvable during this project
Physicians and behavioural scientists	Insufficient knowledge of hearing impairment and treatment	Interactive training, including: - report about experiences in daily practice by a person who is hearing impaired - attractive, compact information material* - emphasizing long term goals on behaviour and quality of life
	Other priorities	Accreditation of the training
Speech and language therapists	Insufficient expertise in adjustment of hearing aids and hearing rehabilitation in this population	Interactive training using practical experiences within the group Attractive, compact information material*
	Lack of time	Discuss with management
Family of participants	In general, little knowledge of hearing impairment and the possibilities for treatment	Information meeting Individual instruction to family by trainer or speech and language therapist Development of information material*
Participants	No available information or instruction material for adults with intellectual disabilities	Development of information booklet adjusted to adults with ID.[14]

* *When this study started, information and communication technology was only just being introduced in Dutch ID centres so advanced ICT techniques were not used in our training and education programme*

3 RESULTS

3.1 Population

Five residential and day-care ID service providers consented to participate: two residential service providers were in a process of developing community based services, two had always been community-based (day-care centres and community homes), whereas the third community-based service, a single group home, offered to participate in a relatively late stage of the study. In five residents of this group home, hearing aids had been prescribed a year earlier, but the prescriptions were not carried out. Three audiological centres were involved in cooperation with these five participating ID service-providers.

Due to delays, we had to stretch the preparatory and intervention phase from 1;3 to 2;3 years. In spite of this prolonged period, realisation of the four modules of audiological rehabilitation could only be undertaken in the two residential ID services. In the two community-based services, preparations had only reached the stage of the consent and inclusion procedure. In the group home that was included at a late stage, an exception was made for the training course: because of the time schedule, carers were trained by the researcher. Acoustics were not assessed.

As a result, within the time frame of the study, we were able to identify 71 hearing impaired persons with mild to severe ID, who were recommended for hearing rehabilitation. Table 3 shows consent and exclusion; we eventually included 31 participants.

Table 3 Consent and inclusion

	n	**n**
Clients with mild to severe intellectual disability and hearing impairment		71
No consent	7	
Consent procedure not completed in time	4	
Consent to participation		60
Exclusion criteria		
formerly prescribed hearing aid	17	
autism/visual impairment	9	
diagnosis of autism/visual impairment not completed	2	
To be included in programme		32
Other reasons for non-participation:		
death	1	
Final number of participants		31

3.2 Encountered barriers

Encountered barriers are presented in relation to the professionals, to the social context, to the organisational context, and to project management.

3.2.1 *Encountered barriers in relation to the professionals*

3.2.1.1 Management

1. Serious delay in decision procedure to participate in the study.
   In one of the ID centres the decision-procedure took 14 months. In another ID centre, senior management indicated that they would decide on the feasibility of the project once the following information was available: numbers of includable participants, necessary financial investment, agreement reached upon the feasibility of the project in the different sections of the organisation that would be involved. This altogether took 16 months.
2. Temporary assignment of both trainers (for the duration of the study).
3. Other priorities.
   Due to other priorities, acoustical adjustments would not be performed within the next year and a half.
   The trainer was withdrawn by management from the training work because of other priorities in her job, no successor was appointed.
4. Limited time of the speech and hearing therapist for hearing aid fitting (two hours a week).
   Due to this participants had to be put on a waiting list. Follow-up of these participants was no longer possible within the time frame of the study.
5. No extra working hours for the coordination of the project.
   Four of the five project coordinators (one speech and hearing, three physicians and one team leader) had to manage the implementation project within their normal schedules.
6. Insufficient expertise in project management.
   None of the coordinators within the ID centres had any expertise in implementing a multidisciplinary protocol; this meant that a lot of active support was required from the researcher, who in turn was not familiar with routines within the ID services.

3.2.1.2 Audiologists

1. Acoustical conditions not assessed by audiologists.
   In spite of the training course for audiologists, one of the audiological centres did not assess acoustic conditions. They did not consider this to be a task for audiologists, because of the required specific expertise that is needed to give a thorough advice on improvements. The other audiological centre did not want to purchase expensive equipment that they considered to be necessary for the assessment. Because of the study, they contracted the assessment to a specialised firm and paid the expenses themselves.

3.2.2 *Encountered barriers in relation to the social context*

1. Poor cooperation between disciplines.
   In one of the ID centres, at a very late stage it became clear that part of the rehabilitation programme could not be realised. It appeared that the necessary steps in the decision procedure had not been ensured. This had remained hidden due to the lack of cooperation between the various parts of the organisation.

3.2.3 *Encountered barriers in relation to the organisational context*

1. Severe delays in audiometry of unknown cause.
2. No speech and hearing therapist available at an ID centre. A speech and hearing therapist in the general community lacked expertise in hearing aid fitting and training.
3. Postponement of fitting with hearing aids in one client due to an imminent change of home.
4. No budget for acoustical assessments.
   One out of the three audiological centres performed assessments, but started to charge a fee during the project once they had gained enough expertise. This could not be afforded by the group home.
5. Some of the clients lived in houses with acceptable acoustical conditions. They would move within months to newly built houses with poor acoustics (decided prior to the diagnosis of hearing impairment).

3.2.4 *Encountered barriers in relation to the project management*

1. Communication problems between an ID centre and the district audiological centre.
2. Severe delays in audiometry were caused due to the fact that an audiological centre and ID centre were waiting for each other to take the initiative for testing.

3.2.5 *Miscellaneous*

1. Technical problems with the portable auditory brainstem response equipment.
2. Vacancy of speech and hearing therapist filled after the training-course.
3. Practical obstacles for acoustical adjustments.
   In the ID centre where acoustical adjustments would be performed, this could not be done within the time frame of the study. This was the result of some practical issues: 1. the client for whom it was performed would move to a another, still unknown location, 2. the ceiling was too low for applying sound absorbing material, and 3. a major renovation was necessary.
4. Organisation of training on too short notice. This was caused by the severe delays in audiometry.

3.3 Encountered facilitating initiatives

The encountered facilitating initiatives are discussed in relation to the professionals and to the social context.

3.3.1 *Encountered facilitating initiatives in relation to the professional*

3.3.1.1 Speech and Hearing therapists

1. The speech and language therapists of one ID centre took initiative to ensure continuity of the habituation training: they convinced management of the need of this and eventually were provided with appropriate time to offer this improvement in audiological rehabilitation to every future client.

2. Speech and language therapists in the one ID centre where carers were trained by the trainer, applied for extension of their work hours to train carers themselves after the trainer, who had a temporary assignment, had gone.
3. A speech and hearing therapist intended to ask for attention for acoustical conditions, if houses belonging to their ID service would be rebuilt or newly built.

3.3.1.2 Audiologists One of the audiologists developed a computational model to assess acoustical conditions which he made available to his colleagues during a special audiologists meeting on acoustics in ID services.

3.3.1.3 The technical service The technical service in one ID centre had first ideas on how to ensure continuity of attention for acoustical conditions.

3.3.2 *Encountered facilitating initiatives in relation to the social context*

3.3.2.1 Communication group In one ID service, the project coordinator became a member of the 'Communication' group to ensure transfer of information from the start to all the staff involved.

3.4 Fulfilment of criteria for completion of the intervention phase in individual participants

Even in the two ID centres where the intervention phase could be completed, the success was variable (table 4). None of the required acoustical adjustments was performed in time and only a limited number of the carers were trained in one ID centre, whereas in the other ID centre no training took place in spite of a local trainer trained by us. 'Communicative intervention' was the only module that was performed in all participants. All the criteria for completion of the intervention phase were met in 3 out of 9 clients from one ID centre, part were met in 28 clients. So we did not even reach the phase in which we could study adoption of the audiological rehabilitation protocol by the participant and disciplines involved.

Table 4 Realisation of audiological rehabilitation in 31 clients

Interventions	**Accomplished**	**Not accomplished**
Hearing aid training	11	20
Advice on communication given	30	1
Training of carers	10	21
Assessment of acoustical conditions	25	6
Performance of necessary acoustical adjustments	0	6

We conclude the following:

1. It turned out to be not feasible to implement an audiological rehabilitation programme in ID centres within a period of 2;3 years.
2. We do realise that the implementation process was seriously hampered by severe delays in audiometry.

3. Barriers were already met at an organisational level of ID services as well as of audiological services before reaching the client-care level where the research group had expected the barriers to occur.
4. Initiatives were taken by professionals to ensure continuation of parts of the rehabilitation programme, which otherwise would not have been the case.
5. Local management of the implementation of a complex multidisciplinary rehabilitation protocol is severely underestimated by management with regards to the required expertise and time.

4 DISCUSSION

This is a report of a failed implementation of an audiological rehabilitation protocol in Dutch intellectual disability (ID) service-providers. It appeared to be not feasible to complete audiological diagnostics and perform all the interventions necessary for an optimal start of fitting with hearing aids within a period of over two years. We had anticipated difficulties, because such rehabilitation requires extra investments, other than in the general population. For instance in order to get the hearing aids accepted, a longer habituation and training period may be necessary, requiring an expert speech and language therapist. Further, specific expertise and continuous support of carers are of paramount importance to make this acceptance last. We had however expected to overcome such obstacles by providing all advice and training in one package and applying a comprehensive implementation strategy, aimed at all involved disciplines, as well as continuous support during the study. Indeed, local coordinators and staff of the participating ID services have put considerable efforts into the project. So what went wrong?

Our implementation strategies focussed primarily on information, training and motivation on the level of professional disciplines within ID services. During the implementation process, it became evident that factors related to the organisation of audiological centres as well as of ID service-providers may have had an important influence. We realise that one unfavourable factor has certainly been the complex nature of the rehabilitation protocol. In the first place, three different types of health service were involved (audiological centres, ID service-providers and specialised institutes for the deaf and hearing impaired). Moreover, the programme involved many disciplines, and several parts of the rehabilitation programme were innovative (on-site audiological assessments, hearing aid habituation and auditory training, assessment and improvement of acoustics, assessment of communication skills and needs by behavioural scientists, training of carers). A third unfavourable factor was that previously, individual hearing assessments were only revealing the tip of the iceberg of hearing impairments in the population with ID. As a result of the current screening programmes, many people with a hearing impairment were now newly diagnosed. It meant a huge additional workload for both audiological centres and ID services. Under-rating the organisational and financial consequences of the project has certainly played a role in the disappointing implementation of our programme. Indeed, considerable financial investments in improvement of unfavourable acoustics, could usually not be realised before the next year, because budgets were to be estimated in the preceding year.

Increasing scientific knowledge on risks, diagnosis and treatment of hearing impairment in persons with IDs puts a claim on ID service-providers to realise their responsibility and invest in specific quality improvement of their care. To our knowledge, no studies have been published on quality improvement of medical care in ID centres. Therefore, our next step was to apply qualitative research techniques to investigate in depth which factors may have positively or negatively influenced the implementation of the audiological rehabilitation programme, in the same three audiological and five ID centres. The results of this investigation have been described in a sequel to this article.

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6 Chapter

IMPROVING THE QUALITY OF AUDIOLOGICAL REHABILITATION IN SERVICES FOR ADULTS WITH INTELLECTUAL DISABILITIES

PART II

FACTORS THAT INFLUENCE THE IMPLEMENTATION

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ABSTRACT

Background:	Although people with an intellectual disability (ID) are at an increased risk of hearing impairment, audiological rehabilitation often fails. In five residential and day-care intellectual disability service-providers, collaborating with three district audiological centres, implementation of an audiological rehabilitation protocol had been unsuccessful, in spite of multiple implementation strategies.
Aim:	To identify and understand factors within ID services and audiological centres that influence implementation of audiological rehabilitation in adults with an ID.
Study design:	Retrospective qualitative study
Method:	Semi-structured interviews of 24 members of staff and management and 3 audiologists. Flaws in implementation strategy/management were identified from the perspective of Grol's implementation model; other factors could be explained from the theory of organisational culture.
Results:	As flaws in implementation strategy/management were identified: insufficient support by staff; insufficient mapping of financial and organisational consequences; insufficient mapping of necessary procedures in ID services; competition by other projects; no strategy for audiological centres; tasks and responsibilities of local project coordinator insufficiently determined; information did insufficiently reach all disciplines involved; insufficient embedding of the project/audiological rehabilitation protocol. The following aspects of organisational culture appeared of influence: quality of transfer of information; focus on quality improvement; focus on results; commitment and responsibility; formal relationships between disciplines in ID centre; policy of ID service-providers concerning normal versus specialised health care.
Recommendations:	Structured collaboration between audiological centres and ID services; hearing aid habituation and training by speech and hearing therapists from audiological centres; systematic training on hearing impairment of professional workers in ID care; obligatory building standards for acoustics in schools, homes and day-care centres; evaluation by ID centres of quality of information streams, effectiveness of interdisciplinary collaboration and decision procedures, as well as actual characteristics of their organisational culture.

A concise report of chapter 5 and 6, titled 'Audiological rehabilitation in adults with intellectual disability: why does it fail?', has been published in the Journal of Policy and Practice in Intellectual Disabilities 2005;2:66-67.

1 INTRODUCTION

This qualitative study is a sequel to a study in which we investigated the implementation of an audiological rehabilitation protocol, adjusted to people with an intellectual disability (ID), in day-care as well as in residential ID services (chapter 5,[1]). The protocol consisted of: 1. hearing aid fitting and training, 2. assessment of and advice on acoustical conditions, 3. behavioural assessment of and advice on communication skills and needs, and 4. training of staff. After all four modules had been arranged by the research team, specific strategies were developed to overcome anticipated barriers, such as a lack of knowledge and motivation of staff. Local project managers were appointed by management of ID centres. A helpdesk was provided for by the researcher. In spite of all this, two of the five participating ID services were not able to complete diagnostic procedures and inclusion of participants within the stretched time frame of the study (2;3 years instead of 1;3 year), whereas the rehabilitation remained incomplete in a majority of participants in the other three services. All interventions that were necessary for an optimal start of the hearing aids could only be realised for 3 out of 31 included persons. Indeed, this implementation failed.

It appeared that problems in ID services already arose at an organisational level before the clients could actually start wearing hearing aids. Practical barriers were identified that caused severe delays or obstructions to the implementation (chapter 5,[1]). Although we could observe which barriers arose, we had insufficient insight into why they arose. Therefore, a retrospective qualitative study was set up to answer the following questions:

1. Which factors influence the implementation of audiological rehabilitation in ID services at an organisational level?
2. Can these factors be explained from flaws in implementation strategies or in project management by ID services?
3. From which theoretical perspective can the remaining factors be understood?

2 PARTICIPANTS AND METHODS

2.1 Participants

Participants were 24 members of management teams and staff of five ID services (2 residential and 3 community-based) and 3 audiologists from three district audiological centres, who had been involved in the implementation of the audiological rehabilitation protocol. The distribution over the disciplines was as follows: 5 senior managers, 7 middle managers, 2 speech and language therapists, 1 trainer of carers, 5 intellectual disability physicians, 2 behavioural scientists, 2 technicians and 3 audiologists. Three senior managers and two physicians who also were project coordinators, belonged to the community-based ID services, and the other interviewees - except for the audiologists - to the two residential ID services.

2.2 Interviews

All participants were interviewed individually by the researcher by means of semi-structured interviews.[2] During and after the interviews, interpretation of information was checked with the interviewees (member check). Topics of the interviews were based on clinical experiences of the research group, as well as on the implementation model of Grol (table 1). This top-down implementation model was chosen, because it also pays attention to bottlenecks for and needs of individual clients and care-providers.[3,4] A constant comparative approach, in which data were collected and analysed concurrently, was adopted. By using this approach, it was possible to incorporate and explore both expected and emergent ideas and themes in subsequent interviews.[5] The interviews lasted between 60 and 90 minutes each and were recorded on tape after consent of the interviewee. The taped interviews were transcribed literally by an independent co-worker.

Table 1 Implementation model of Grol[3,4]

- *A Implementability of an innovation: estimate the chances of a successful implementation*
    - determine support
    - determine relevant features of the innovation
    - adjust innovation to wishes of consumers
- *B Diagnosis of the situation: identify factors that influence implementation*
    - determine interests and objectives of people involved
    - describe present procedure(s)
    - determine hampering and stimulating factors
    - distinguish subgroups in the target group
- *C Implementation strategy: set up a programme for implementation*
    - choose one or more strategies
    - make a concrete plan of approach and carry this out
    - take care of ensuring the accomplished change
- *D Evaluation: evaluate progress and effects of the implementation and adjust the plan if necessary*
    - evaluate effects
    - evaluate progress of implementation
    - evaluate costs in relation to effects

2.3 Analysis

The transcripts were read and re-read and the data organised under initial, administrative codes. By constant comparison of coded fragments on similarities and differences, themes were identified that were relevant to the research questions. During the interviewing period, this process was repeatedly and randomly checked by a peer group consisting of two colleagues. Differences in interpretation were discussed and adjustments were made, until agreement was reached.

The implementation model of Grol (table 1) was used again as a structural basis to check, which themes could be explained by shortcomings in our implementation strategies or in local project management in the ID services. All fragments were reread from the perspective of this model, and additional themes were identified.

Relevant factors which could not be explained from this viewpoint, mostly concerned attitudes of staff and management. The theory of 'organisational culture',[6] appeared to offer a background from which these fragments could be explained. Again, the fragments were reread and additional themes identified from the perspective of organisational culture.

3 RESULTS

Eight themes were identified, important with regards to flaws in implementation strategies or local project management, as well as six themes related to organisational culture, which had positively or negatively influenced implementation of the audiological rehabilitation protocol.

3.1 The implementation strategy or local project management

Themes concerning the implementation strategy or local project management are shown in table 2, and some examples of quotations of the interviewees are presented in box 1.

Table 2 Flaws in implementation strategy or local project management

A *Implementability of the innovation*
    1. Insufficient support by staff
    2. Insufficient mapping of financial and organisational consequences

B *Diagnosis of the situation*
    3. Insufficient mapping of necessary procedures in ID services
    4. Competition by other projects in ID services

C *Implementation strategy*
    5. No strategy for audiological centres
    6. Tasks, responsibilities and coordination insufficiently determined
    7. Information insufficiently reached all disciplines involved
    8. Insufficient embedding of the project or audiological rehabilitation protocol

We concluded that:

1. a more detailed description of financial and organisational consequences of participation in the project (theme 2) should have been provided earlier in the project
2. the local technical and organisational procedures (theme 3) were too different between institutions. Procedures were also complex, as is shown in table 3 in which the many people are shown who were involved in technical acoustical adjustments in one of the ID services. An inventory of this before the project started should have been made with subsequent adaptations to the implementation plans accordingly.
3. given the aforementioned, more time should be made available to influence management structures.

4. an inventory should have been made of barriers in the audiological centres too, with subsequent strategies to overcome them (theme 5). The sudden increased stream of patients was a cause of capacity problems for these centres. We had insufficiently anticipated the lack of experience with performance of on-site audiometry in the ID services and the underrated amount of extra time and manpower needed. After our specific training, only one audiological centre provided on-site assessment of the acoustics; the second centre contracted this to a commercial firm, they did not want to purchase the expensive equipment that they considered to be necessary for the assessment and did not feel expert enough to give advice on necessary improvements, and the third audiological centre considered this not to be a task for audiological centres despite earlier discussions.
5. tasks, responsibilities and coordination of activities of both ID services and audiological centres (theme 6) should have been made clear earlier in the project.

Table 3 Route of the advice for technical acoustical adjustments in one of the residential ID services

Audiological centre: assessment of acoustics → advice for technical acoustical adjustments → referring physician → personal carer of client → assessment of priority by team of carers → assessment in relation to budget by middle management → technical service (< --- > contractor for estimation of costs) → assessment of priority by senior management → technical service → contractor

Themes 1 (insufficient support by staff), 4 (competition by other projects), 6 (tasks, responsibilities and coordination insufficiently determined), 7 (information insufficiently reached all disciplines involved) and 8 (insufficient embedding of the project or audiological rehabilitation protocol) could only have been anticipated and handled by local project managers, familiar with the organisational structure of the ID centre, and with enough capacities and support to develop strategies to foresee and overcome such barriers.

3.2 Aspects of organisational culture

Apart from strategic flaws, the following aspects of organisational culture within ID services were identified, which may have influenced the implementation: 1. quality of transfer of information, 2. focus on quality improvement, 3. focus on results, 4. commitment and responsibility, 5. formal relationships between services in the ID centre, and 6. policy of the ID service concerning normal versus specialised health care (quotations in box 2). Themes 1 and 6 emerged from interviews in all five participating ID services, whereas the other four aspects originate from the interviews in the two residential ID services where the entire intervention phase for the implementation could be completed.

The quality of transfer of information (theme 1) was reported to be poor in both residential and community-based services. It concerned several professional disciplines and written as well as oral information (table 4).

Box 1 Flaws in implementation strategy/management: quotations from interviews

1. Insufficient support by staff

"If my grandfather gets a hearing aid, my whole family wouldn't have to follow a training course?" (carer to trainer)

"If I talk to him, he understands me. He doesn't want to hear, he is just playing deaf." (carer to physician)

"In a home for clients with severe behavioural problems, carers are afraid that if the clients would hear too much, they would get too many stimuli, resulting in more behavioural problems." (physician/project coordinator)

"If middle managers commit themselves to the project, then they will be more inclined to say to carers: "Listen, we have agreed upon that, you will have to do it", whereas now it has been a top-down decision, and then it depends on the individual motivation of cluster managers how they address the carers." (physician)

2. Insufficient mapping of financial and organisational consequences

"It is important to get the organisational part more clear before the start of a project, because our organisation is not equipped to do this." (physician)

3. Insufficient mapping of necessary procedures

"The technical service already said at the moment we sent them the reports, that it would be a problem to realise it (i.e. adjustment of acoustical conditions) at short notice, because next year's budget was already decided on." (audiologist)

4. Competition by other projects

"We have to pull out all the stops to perform our daily care and we can't have all this added to it." (carer to physician)

"We are very ambitious and innovative and because of that there's a lot of enthusiasm for new things and projects and so on. But they forget we are working on 20 plans, while in the organisation there's only room for ten. And then you perform 20 plans poorly in stead of 10 well." (middle manager)

5. No strategy for audiological centres

"What turned out to be more troublesome than we thought, was the manpower we had to put in and the fact that we were unfamiliar with this way of working (i.e. on-site). We had to find our way. During the process, bit by bit agreements on the approach were made with the intellectual disability services." (audiologist)

"The moment the clients would actually receive their hearing aids, has eventually been totally unclear to us (i.e. the audiological centre), and therefore no appointments have yet been made for the checks on the hearing aids. This would not have happened with a proper coordination." (audiologist)

6. Tasks and responsibilities of local project coordinator insufficiently determined

"It wasn't that it was all worked out what I had to do in those four hours, it was more like what I needed to do for you (i.e. the researcher)." (project coordinator/speech and language therapist)

"It was more like I should play a central part in it, function as a sort of go-between, that's about it." (project coordinator/physician)

7. Information insufficiently reached all disciplines involved

"It was noticed that when a client was invited for the check up of earwax or the audiometry, carers always asked: what's going on, what do we have to do, is this necessary? As though people didn't know what was going on, never had heard of the study." (physician)

"Provision with information of carers, one should specifically reflect on that, not just sending information to the middle management, but to invest more intensively in informing people on the work floor." (speech and language therapist/project coordinator)

8. Insufficient embedding of the project or audiological rehabilitation protocol

"It is very important not only to start new things, but also to implement them properly in the organisation, to evaluate them well, to come to an agreement on responsibilities, to round it off and not to let it die a silent death, which happens regularly." (middle manager)

"In a lot of ID services it's a problem to secure continuity of appointments for a client in the future." (audiologist)

Table 4 Poor transfer of information

- poor transfer of client information to audiological centres
- appointments for audiometry insufficiently passed down within teams of carers
- poor feedback of audiometry results by physicians to (family of) participants, speech and language therapists and carers
- poor quality of transfer of information within teams of carers
- written information is hardly read by carers
- insufficient responsibility to take care of messages for absent colleagues
- insufficient information by project coordinator towards the management team

Focus on quality improvement (theme 2) was a central reason for management of ID services to participate in the study. It also influenced the receptiveness of carers for new information: some were open to innovations that might improve the quality of life for their clients, whereas other carers from the same ID service evidently were not. Quality improvement was the main drive for speech and language therapists in one ID service to develop an adjusted hearing aid training, to be offered to future clients.

A senior manager suggested that the relatively successful implementation in his ID service might be due to a policy of 'keeping promises', which was part of a management strategy, focussing on results (theme 3). He explained that since a few years, organisational culture had been explicitly changed into this direction, in contrast with a less critical attitude in the preceding years, because of increasing demands upon quality of care by health authorities as well as clients or their legal representatives. For the current project, this policy had resulted in recruitment of sponsors for the training of carers, and in flexibility with formal procedures for renovations of homes.

Commitment and responsibility (theme 4) influenced the implementation process in a positive way. It became visible in the attitude of staff: positively in initiatives of carers to ensure the implementation in daily life by means of concrete agreements, negatively in neglecting maintenance of hearing aids (rusty batteries) or no replacement of lost hearing aids. A positive experience was, that when the trainer fell ill in one of the ID services, the speech and language therapists intensified their instructions to carers and developed a project to continue the training of carers in the future.

Effects of different formal relationships between disciplines (theme 5) became also apparent. In one residential ID service, a middle manager reported that although physicians professionally advised on aspects of healthcare (i.e. training of carers and acoustical adjustments), managers of homes, who controlled the budget - and might have other priorities - decided whether improvements were actually carried out. Indeed, the physician/project coordinator in question remarked, that he had no influence on the decision procedure within homes. In contrast, in the other residential ID service, the implementation project was embedded in a product group 'Communication', to ensure a good communication between all disciplines involved.

Box 2 Aspects of organisational culture: quotations from interviews

1. Quality of transfer of information

"In daily practice you notice quite regularly, that what you have said is handed over differently or is reported differently, if it is reported at all, or that the carer's own opinion is added to it." (physician/project coordinator)

"Or someone is on a holiday or prolonged absence by illness, and nobody sees to it that somebody else takes over (i.e. checking the mail)." (behavioural therapist)

"Communication in our organisation is lousy. I think that's why a lot of these projects fail. ….. It's because it is too big, not manageable. I think this is in fact the central issue why things go wrong." (physician/project coordinator)

2. Focus on quality improvement

"We profited from our training course, because we are developing a training course for clients, with which we can work a lot more secure. We want to accompany all new clients who get hearing aids in this way." (speech and language therapist)

"I think it's very individually coloured by the employee involved: I know of employees that embrace new ideas, almost run away with it, whereas other people don't do this." (behavioural therapist/middle manager)

3. Focus on results

"If we focus on a possible factor of success in this project - and really, I still think it could have been a lot better - but at the moment a unit decides to participate in the project, then there can't be any 'yes, but's afterwards." (senior manager)

"Instead of following the normal procedure, because of the high priority, it was directly put on the list of projects to carry out, and we said, we will finance this one way or another." (technical service)

4. Commitment and responsibility

"This carer's attitude was something like: we have to make concrete agreements on this, and properly disseminate information, and everybody has to stick to it, so she is very committed now." (speech and language therapist/project coordinator)

"I had the feeling no one in our organisation felt responsible for the overall picture." (behavioural therapist)

"There are clients who have lost their hearing aids half a year ago, and that is mostly due to inattentiveness of carers who have let it slip." (physician/project coordinator)

5. Formal relationships between disciplines in the ID centre

"I think, too many people are involved. Choices made by someone with respect to the content, are eventually decided upon by someone else who controls the budget and who has other interests." (middle manager)

"With regard to the training of carers and acoustical adaptations, you deal with other departments, in which you have no participation, in which one is dependent on other persons' goodwill, and then it's pretty rotten to experience that it fails." (physician/project coordinator)

6. Normal versus specialised healthcare

"There are still too many people who say: community-based care means that you have to get rid of all specific expertise, which should be integrated in general health care." (senior manager)

"Beforehand it is not taken into account, that clients can suffer from hearing impairments; social issues get more attention. Focus on the health status of a client, including hearing impairment, comes second if not third." (senior manager)

Policies of ID services concerning normal versus specialised healthcare (theme 6) also appeared to have an influence. In the Netherlands, as in other countries, there is a strong tendency for adults with an ID towards a normal life, integrated in the community. In two out of the three participating community-based organisations, this was interpreted as 'clients should use the same healthcare services as everyone'. As a result, no specifically experienced speech and language therapist was available for these clients to provide hearing aid habituation and training. A senior manager of one of these ID services remarked that social issues (work, social activities) were given more attention than medical issues like hearing problems. The transition of culture in ID care had also led to a social-educational instead of a nursing background of carers in all participating ID services. As a result, much time had to be invested into repeated information of carers.

4 DISCUSSION

Summarising the issues, we find first explanations for the fact that, although hearing impairment is a highly prevalent co-morbid condition in adults with intellectual disabilities (ID), audiological diagnosis and rehabilitation frequently fail in this group. We showed in a first study, that this failure is not in the first place the result of a lack of understanding or cooperation by these people, as is usually supposed by physicians (chapter 5[1]). Main reasons are practical barriers in regular audiological care on the one hand, and the dependence of persons with ID on other people and on ID services on the other. Unfavourable acoustics, specifically in modern group homes and day-care centres, require expert on-site judgement and financial investments, which are both difficult to acquire.

This study shows that, although most regional Dutch audiological centres have the necessary technological expertise and equipment to diagnose and advise difficult-to-test clients, and most are now aware of this specific risk group and prepared to accommodate it, the sudden increased stream of patients, and the on-site audiometry in the ID centres caused capacity problems. Judgment of communication capacities by behavioral experts and effective information of caregivers, which are offered to young Dutch children with a severe hearing impairment or deafness by audiological centres or by specific family guidance services, are not available to adults with an ID. On-site judgement of acoustics is innovative, and requires specific training as well as extra budget. Apart from this, not all audiological centres consider this to be their task. In our implementation strategy, we had not anticipated such problems in audiological centres, also because of the discussions at the level of the Federation of Dutch Audiological centres. These problems led to major delays in diagnostic procedures and hearing aid prescriptions.

In ID services, other problems were met. When we approached residential and day-care ID services to participate in the programme, the management just started to realise that hearing impairment among their clients might be a point of attention. Nevertheless, senior managers of five ID services were prepared to invest in a rehabilitation programme, aiming to improve their quality of care. The failing implementation was partially caused by the aforementioned delays in diagnostic procedures, leading to a lack of time for ID services to properly organise trainings, reserve budget and start procedures for technical adaptations, which also took more time than the research group had taken into account. Further, this study confirms that there is a lack of knowledge on hearing impairment among professional disciplines in ID services, with the result that hearing impairment is not always recognised in clients, or consequences are underrated and because of this, treatment not supported. Moreover, the transfer of professional information between and within disciplines, as well as between audiological centres and ID services, appeared to be poor.

Our preparations of the implementation had insufficiently taken into account details of the organisational and financial consequences of participation, and formal local procedures in ID services. This resulted in other unanticipated obstacles to the implementation of the rehabilitation protocol. Apart from these flaws in our implementation strategy, it was apparent that the research group, as well as the management of ID services, have underrated the workload for and necessary expertise of the local project coordinators. These should ideally be able to monitor information and support during the implementation process, to develop strategies if needed, to adjust to competing priorities, and to ensure continuity after the study has ended. For this task, professionals like physicians or speech and language therapists may not be sufficiently equipped.

In conclusion, the combination of all these factors - the innovative and complicated rehabilitation programme, the lack of capacity and means of audiological centres to effectively accommodate this new patient group, the introduction of innovative expertise on acoustics, insufficient knowledge on hearing impairment of professionals in ID services, a failing transfer of professional information between audiological and ID centres as well as within ID services, the requirement of considerable financial investments and flexible local procedures, and local project managers who were insufficiently equipped for their complicated task - offer more than enough explanation for the failing implementation. Failing rehabilitations prior to the programme are no longer surprising.

Aspects of organisational culture, reflected in the behaviour of staff and management,[7] may also have influenced the implementation process. Since we were only able to retrospectively study such aspects in two residential ID services, these findings are to be considered with prudence. Nevertheless, they are a first tentative inventory of such aspects in ID services. For indeed, studies of the relationship between successful implementation of medical improvements and organisational culture in healthcare have been published before,[8,9] but not for multidisciplinary medical improvements in ID care. Quinn and Kimberley[10] have defined four types of organisational culture: group culture (key features: teamwork, commitment, participation), developmental culture (risk taking, innovation, creativity), hierarchical culture (formalised, procedures) and rational culture (focus on results, efficiency). Shortell et al[11] have described a positive relationship between implementation of continuous quality improvement in hospitals, and cultures that can be identified as group and developmental cultures. The themes, identified in our study, suggest that this might be similar in ID services: 'commitment and responsibility' may indicate aspects of group culture, whereas in 'focus on quality improvement' the positive influence is shown of an aspect of developmental culture. On the other hand, 'formal relationships between disciplines' may reflect remnants of the hierarchical culture in the former institutions. The theme 'focus on results' was mentioned by a senior manager as a possible explanation for the relative successful implementation of the protocol in his ID service. 'Focus on results' is an aspect of a rational organisational culture, which is considered more or less natural in hospitals. We might hypothesise that in ID services, a rational culture is less self-evident, reflecting the chronic character of this type of care, the lack of demands by clients and their family, and the fact that community living, work and social activities may be given a higher priority than diagnosis and treatment of co-morbid conditions.

Our study has been performed in the Netherlands, but we do not think that the results reflect difficulties that do not exist in audiological care for persons with IDs in other countries. However in several countries, as in Norway and Sweden, ID services have been community based for a longer time than in the Netherlands. Hierarchical cultures will probably have disappeared completely, and may have been replaced by the commitment and responsibility, which is often found in small teams. But are such teams equipped to focus on quality improvement and on results? Is interdisciplinary communication effective? Is their knowledge of sensory impairments better than that of their Dutch colleagues? We guess that these questions may not always be answered positively. Indeed in our study, a former institute performed more effective than the three ID services that had always been community-based.

Therefore, based on this evaluation, first international recommendations can be formulated to help improving the quality of audiological rehabilitation in people with intellectual disabilities:

1. Structured collaboration between audiological services and ID services; hearing aid habituation and training by speech and hearing therapists should be performed by audiological centres.
2. Systematic inclusion of education on hearing impairment in basic and postgraduate training programmes for professional workers in ID care.
3. Obligatory building standards for acoustics in schools, homes and day-care centres for people with ID, taking hearing impairments specifically into account.
4. Evaluation by ID services of their responsibilities with respect to physical impairments, the quality of their internal and external information streams, effectiveness of their interdisciplinary collaboration and procedures with respect to audiological rehabilitation, and consideration of actual characteristics of their organisational culture.

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7 Chapter

GENERAL DISCUSSION

INTRODUCTION

In the first part of this thesis, results are presented of a cross-sectional epidemiological study in a population, representative of Dutch adult users of intellectual disability (ID) services. Prevalences of hearing impairment, as well as of dual sensory impairment were estimated and their relation to risk factors. The second part of this thesis focuses on audiological rehabilitation. At first results are described of an explorative pilot-study in which expectations of, and satisfaction with hearing aids are studied in adults with an intellectual disability (ID). This is followed by results of a study in which we investigated the feasibility of the implementation of an audiological rehabilitation protocol in Dutch ID service-providers. The implementation process was evaluated by means of qualitative research methods. Factors are presented that influenced the implementation process. In this final chapter we will comment on the study results and on the problems that were met during the studies. We will offer recommendations for existing guidelines and for the organisation of audiological care for adults with an ID, and offer suggestions for future research.

THE SIZE OF THE HEARING PROBLEM

Our epidemiological study was the first nation-wide study in adult users of residential and day-care intellectual disability service-providers, applying generally accepted diagnostic methods and WHO-criteria for hearing impairment and the classification of its degree. We have shown (**chapter 2**) that with a re-weighted prevalence of 30.3%, hearing impairment occurs more frequently in this population compared with the prevalence of 16 - 17% in the general adult population.[1,2,3]

Prior to the study, we wondered whether the increased risk of hearing impairment in the population with an ID could entirely be explained by risk factors as age and Down syndrome. We could however also demonstrate a significantly increased prevalence of hearing impairment in the subpopulation with an ID by a different etiology than Down syndrome as compared with the general population. This significantly increased prevalence appeared to occur in all age groups from 18 - 30 years and onwards. As a result, the prevalence-versus-age curve was shifted forwards with one decade (figure 2, **chapter 2**). As opposed to the population with Down syndrome, we do not a priori expect premature ageing on top of the congenital or early childhood impairments in this group. Conductive losses might partially explain the increased risk. Our data enable further analysis of the type of hearing impairment, but this could not be realised within the time frame of this thesis. We therefore do not know to what extent conductive losses are responsible for the hearing impairment in this subpopulation. It may very well be possible that middle ear pathology quite often remains undetected, because of a lack of complaints, or misinterpretation of symptoms by carers. Untreated chronic middle ear infection may secondarily lead to chronic perforation of the tympanic membrane or cholesteatoma,[4] but also to sensorineural hearing loss.[5,6] Our hypothesis of unidentified middle ear pathology is supported by results of a study of

Evenhuis et al[7] in which an unexpected amount of conductive losses were diagnosed in elderly people with an ID by other causes than Down syndrome.

The shifted prevalence-versus-age curve leads us to recommend an adjustment of the Dutch and IASSID consensus guidelines for hearing screening in adults with an ID[8] by proposing to shift screening of age-related losses forward by 10 years (from 50 to 40 years) for persons with an ID by other causes than Down syndrome

With an odds ratio of 5.18, Down syndrome could be identified as the most important risk factor for hearing impairment in the adult population with an ID. The hearing impairment in adults with Down syndrome may be of a conductive and/or sensorineural nature. In **chapter 2** we discussed that the conductive component may be caused by congenital malformations of the middle ear, by frequently occurring or chronic middle ear infections persisting into adulthood, or by age related sensorineural hearing impairment, which already occurs during the second decade of life.[9]

We wonder if rigorous treatment of middle ear infections from childhood in people with Down syndrome, will reduce the very high prevalence of hearing impairment (reaching 100% in persons over 60 years) in the long run. This effect has already been shown for the short term by Shott et al.[10] Forty-eight children with Down syndrome under the age of two years received aggressive 'state of the art' treatment in case of otolaryngologic problems and were followed up during a period of five years. Prior to treatment, 81% of the children had abnormal hearing levels, ranging from borderline normal or mild loss to severe loss. After treatment with antibiotics or (repeated) ventilation tubes, 97.7% of the children had (borderline) normal hearing evaluations and 2.3% mildly abnormal hearing. Future research is needed to investigate the long term effect of rigorous treatment of middle ear infections on prevalence of hearing impairment.

Because of the very high prevalences of hearing impairment in adults with Down syndrome, we make a second recommendation for adjustment of the aforementioned IASSID consensus guideline, namely to perform complete audiometry, instead of a hearing screen, every 3 years throughout life in persons with Down syndrome.

We could not demonstrate the strong positive relationship between a more severe degree of ID and hearing impairment which has been shown in the same study population for visual impairment.[11] This difference might hypothetically be explained by two reasons. Automutilation may play a larger role as a cause for visual impairment than for hearing impairment, because the eyes are more exposed. However, we speculate that cerebral visual impairment might be a more important explanation. Visual impairment caused by brain damage is now the leading cause of childhood visual impairment in industrial countries,[12] Periventricular white matter damage (leukomalacia) in pre-term children and neonatal hypoxia-ischemia in at term children are its most common causes.[13] These causes usually lead to other neurological impairments as well, such as cerebral palsy, epilepsy and intellectual disabilities. In consequence, the most important at risk groups are pre-term children, and children with cerebral palsy, especially when combined with intellectual disabilities and epilepsy. In our own population-based study in adults with intellectual disabilities,[14] as well as in the Danish study by Warburg,[15] cerebral visual impairment was diagnosed in respectively 12.6% and 9.6% of the participants.

The auditory tract differs from the visual pathways in such a way, that it contains many alternate pathways from the brainstem to the cortex. As a result, the auditory system past this level seldom breaks down completely. However, hearing impairment may also be caused by pathology in the tract from the cochlea (from the inner hear cells) through the auditory nerve up to the brainstem. In this case otoacoustic emissions, which are produced by the outer hair cells in the cochlea, may still be present. This type of hearing impairment will therefore not be detected when otoacoustic emissions screening equipment is used. However, because in auditory neuropathy otoacoustic emissions eventually may also disappear, we may not have missed all such cases.

THE SIZE OF THE COMBINED HEARING AND VISUAL PROBLEM

Given the increased prevalences of hearing and visual impairment in adult ID service users, we also expected an increased prevalence of dual sensory impairment, as compared with the general population. In this study it was quantified for the first time (**chapter 3**). For the total adult ID population a re-weighted prevalence of 5.0% could be estimated, for the population younger than 50 years of 2.9%, and for the population of 50 years and over of 11.0%. The re-weighted prevalence in the total population, as well as in both subpopulations under and over 50 years were considerably higher than in the general population. A more severe degree of ID, age 50 years and over, and Down syndrome were identified as risk factors. We hoped to get some further insight into the possible causes of dual sensory impairment by studying the aetiology of the ID in the group with dual sensory impairment. This appeared to be unsuccessful, because apart from Down syndrome, which already was identified as a risk factor, the aetiology of the ID had been diagnosed in a minority of cases.

We have shown that dual sensory impairment frequently occurs in adults with an ID, but also that with the present organisation of care, it had not been recognised prior to the study in 88.4% of the cases. Even if treatment or rehabilitation is not possible or accepted, it is important for caregivers to be informed about the dual sensory impairment, as they will have to take this into account in their care. For instance, if surrounding events are insufficiently seen or heard, a more active approach may be necessary to keep clients involved in social events. And when in case of hearing impairment pictures, pictograms or signing are used as additional modes of communication, this is inadequate as the person addressed has also impaired vision. Pictures and signing may need to be enlarged or made more contrasty in order to be properly seen.

On the other hand it is important to realise that only one out of 77 persons was functionally deaf and blind, and two persons were blind in combination with severe hearing impairment. So the severity of dual sensory impairment in a majority of cases permits effective treatment or rehabilitation.

THE SCREENING PROTOCOL

In the Netherlands, first consensus guidelines were published for identification and diagnosis of hearing impairment in children and adults with an intellectual disability, recommending active screening.[16] After this, Dutch physicians working in intellectual disability care took the initiative to organise screens, whereas several specialised organisations for hearing care started in ID centres a system of large-scale screen with otoacoustic emissions (OAE) and impedance audiometry.
The introduction of hearing screening equipment based on otoacoustic emissions during the 1990s has been a big step forward because no active cooperation of the client is necessary. During our preparations, we globally expected 2/3 of the study population to pass, and 1/3 to fail for one or both ears, requiring further audiometry. In fact, only 30% passed the test. A screening method, requiring further assessment in 70%, seems to be not very effective. However, the prevalence of hearing impairment in the population with an ID appeared to be 30%. This means that hearing impairment will be diagnosed in one out of every two clients who are referred for audiometry, which is rather effective. If sedation is necessary for hearing screening, direct referral to an audiological centre for complete audiometry would be a better option.

It needs to be assessed if Automated Auditory Brainstem Response (A-ABR) screening equipment is a better alternative for hearing screening based on otoacoustic emissions. A-ABR is less sensitive for environmental noise and for middle ear problems, and thus might even provide a lesser amount of referrals. With the A-ABR also auditory neuropathy (see also the discussion of chapter 2) will be detected. At present however, A-ABR equipment is not available for persons over six months of age. In addition, this screening method is a lot more expensive than OAE screening.

THE DIAGNOSTIC PROTOCOL

The diagnosis of hearing impairment has long been difficult in persons who were not able to cooperate sufficiently, as in persons with a severe degree of ID. Since the introduction of Auditory Brainstem Response (ABR) audiometry, in principle hearing impairment can now be identified and quantified in every person who is difficult-to-test. However, although no active cooperation is required with this method, the client should sit or lie quietly and should not make too many noises for about 30 - 60 minutes. Therefore, in anxious or restless clients, sedation may be required, and in a limited number of cases even general anaesthesia. Based on the international guidelines,[8] in our study ABR was recommended for clients who could insufficiently cooperate with pure tone audiometry.

During the last decades, ABR has been applied in Dutch audiological centres mostly to diagnose retrocochlear pathology and much less frequent to assess the hearing threshold in difficult to test patients. Lately ABR is also increasingly used to estimate hearing thresholds in neonates and very young children, and also to estimate hearing thresholds in people with an ID in whom pure tone audiometry appeared to be less feasible. Nevertheless, in spite of our diagnostic protocol, it appeared that in a large majority of the participants observational audiometry had been used instead of ABR because of a reluctance of audiologists to advise sedation, a preference for observational audiometry, or anticipated difficulties to interpret ABR in this group.

As a comparison, in an older institute-based study of hearing function,[17] on-site ABR was applied by an experienced audiological assistant from a nearby academic audiology department in 145/672 study participants. A severe or profound ID was present in 142 cases. It appeared to be feasible to obtain reliable results in 65.5%, and moderately reliable results in 21% of the 145 cases (Evenhuis, personal communication). Oral sedation was necessary in 55%, whereas in 30% repeated appointments were required e.g. because of necessary sedation, occluding ear wax or insufficiently interpretable results. These findings confirm that, even with objective methods, it can be difficult to obtain reliable audiometric data in persons with intellectual disabilities (sedation, repeated appointments), but also that with on-site testing by an experienced professional, data relevant to treatment were obtained in the majority of these difficult-to-test persons.

DEPENDENCE ON CARERS

We studied the expectations of adults with an ID towards hearing aids, and also their experiences half a year after the introduction of the devices (**chapter 4**). In the results, dependence on carers was noticeable in a negative way: information on the hearing aids was incomplete or incorrect in the majority of participations prior to the interviews, complaints of problems with the hearing aids still existed in six out of nine clients half a year after the introduction of the hearing aids.

People with an ID are not the only persons who may have to depend on carers for the use and maintenance of their hearing aids. This is also reported for residents of nursing homes. Cohen-Mansfield and Taylor[18,19] studied barriers to effective utilisation of hearing aids in 33 residents of nursing homes. Most residents (86%) needed help in taking care of the hearing aids, which is comparable with findings in the adults with a mild to moderate ID from our study population. Twenty-three nursing home residents reported to have one or more problems with the devices, e.g. dysfunctioning hearing aids or discomfort. It turned out that staff often did not know why hearing aids were not used, whereas the residents themselves were much more aware of these reasons. In the nursing home however, staff had not been trained in the use and maintenance of the hearing aids, including trouble shooting whereas we did train caring staff. We conclude that in addition to training of caregivers, also a regular check-up of complaints and hearing aids is necessary in adults with an ID by a professional (speech and hearing therapist, hearing aid supplier). This check-up should also be available for persons with a mild ID.

AUDIOLOGICAL CARE

We have described the barriers that were met during the implementation of an audiological rehabilitation protocol, adjusted to adults with an ID, in ID service-providers (**chapter 5**). These barriers were already met at an organisational level prior to the actual introduction of the hearing aids to the participants. We have explained that just fitting of hearing aids is usually not effective in persons with ID, and that training of carers and optimisation of acoustical conditions are indispensable for permanent acceptation of the devices. In spite of specific strategies that were developed to overcome anticipated barriers, such interventions could be realised in only 3 out of 31 included participants within the time frame of the study (2;3 years).

Procedures for training of carers and technical acoustical adjustments appeared to take a long time. This may be caused by the decision procedure concerning priorities in training, or priorities in technical adjustments in the houses, as for instance is shown in **chapter 6** table 3, but also by the fact that training programmes for carers, and necessary adjustments in housing conditions are usually planned a year in advance. In addition, expertise on the use and maintenance of hearing aids, and adjustment of acoustical conditions were not given high priority (**chapter 5**). Our study results indicate that another approach is necessary to provide optimal conditions for fitting with hearing aids through ID service-providers.

We therefore recommend that education on hearing impairment is systematically included in basic and postgraduate training programmes for professional workers in ID care. With regards to the acoustical conditions we recommend that existing building standards for acoustics in schools, homes and day-care centres for people with ID, which take the necessary acoustical conditions for hearing impairment into account,[20] are made obligatory by the government.

It also appeared from our study results that structured collaboration between audiological centres and ID centres is necessary. According to the rehabilitation protocol, audiological diagnostic and rehabilitation for adults with a more severe degree of ID should preferably take place within the ID centre. We further recommend that local 'hearing teams' are formed to provide the necessary care in the ID centres.

These teams should consist of:

- an audiologist: for the introduction and follow-up of the audiological rehabilitation
- a speech and hearing therapist: for the habituation and auditory training, provision of additional modes of communication, regular check-up of complaints by the client and his or her hearing aids
- a hearing aid supplier

- a behavioural scientist in cases with behavioural problems
- a physician for persons with an ID: for the coordination and continuity of the rehabilitation activities within the organisation and with the external members of the hearing team, for a regular check-up of ears to detect ear wax or middle ear infections, and to convince management of the necessity of the fitting with hearing aids including the necessary conditions as described in **chapter 5.**

The meetings of the hearing team should take place at a regular interval. During the meetings the personal carer of the client involved should be present to provide and receive the necessary information. It is optional for the client to be present during the meetings. He or she can also discuss (if possible) his wishes, expectations and experiences with the personal carer and/or the speech and hearing aid therapist.

This proposed structure for collaboration has already been described by Kingma et al in a Dutch ID centre where it operates successfully.[21]

As hearing impairment is present in one out of three adults with an ID, not related to the severity of ID in the population with an ID by a different cause than Down syndrome, service-providers will have to take responsibility and provide the conditions for an adequate audiological rehabilitation, including optimal acoustic conditions in all settings where persons with an ID live and work.

RECOMMENDATIONS FOR GUIDELINES AND AUDIOLOGICAL CARE

In this paragraph we summarise the recommendations for adjustment of existing guidelines and for the organisation of audiological care for adults with an ID, which we offered on the preceding pages.

We recommend:

1. the following adjustments of the existing Dutch and IASSID consensus guideline for hearing screening in adults with an ID:[16,8]
    - to shift the screening of age-related losses forward by 10 years (from 50 to 40 years) for persons with an ID by other causes than Down syndrome
    - to perform in persons with Down syndrome complete audiometry instead of hearing screening every 3 years throughout life
2. that apart from the training of carers in the use and maintenance of hearing aids, a professional help (hearing aid supplier, speech and hearing therapist) performs a regular check-up of complaints and hearing aids.
3. obligatory building standards for acoustics in schools, homes and day-care centres for people with ID, taking hearing impairments specifically into account.
4. systematic inclusion of education on hearing impairment in basic and postgraduate training programmes for professional workers in ID care.
5. structured collaboration between audiological services and ID services by the formation of a hearing team consisting of an audiologist, a speech and language therapist, a hearing aid supplier, a behavioural scientist in case of behavioural problems and a physician for people with an ID or general practitioner.

In addition the recommendations are summarised which were discussed in **chapter 6**:

6. We recommend evaluation by ID services of their responsibilities with respect to physical impairments, the quality of their internal and external information streams, effectiveness of their interdisciplinary collaboration and procedures with respect to audiological rehabilitation, as well as consideration of actual characteristics of their organisational culture.

RECOMMENDATIONS FOR FUTURE RESEARCH

We recommend to study:

- effects of rigorous treatment of hearing impairment in children and adults with Down syndrome on the prevalence of hearing impairment in the long term.
- prevalence and risk factors of auditory neuropathy in the population with an ID.
- whether screening with A-ABR equipment reduces the amount of referrals to audiological centres.
- the cause(s) of the increased prevalence in the subpopulation with an ID by other aetiology than Down syndrome.
- prevalence of hearing impairment in children with an ID.
- effects of audiological rehabilitation.

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8 Chapter

SUMMARY

SAMENVATTING

SUMMARY

In this thesis, the results are presented of an epidemiological study of hearing impairment in adults with an intellectual disability (ID), as well as the results of a study on the implementation of an audiological rehabilitation protocol in centres for persons with an ID. It is preceded in **chapter 1** by a review of literature on epidemiology and rehabilitation of hearing impairment in persons with an ID. Moreover in this chapter is explained why these studies were established. In addition, aims of the study are formulated, and definitions given for intellectual disability, hearing impairment, visual impairment, dual sensory impairment and implementation.

The few studies on prevalence of hearing impairment in adults with an ID, that thus far have been published, were performed in small or selected populations. In addition, different definitions of hearing impairment were used. In order to be able to estimate prevalence and severity of hearing impairment on a nationwide level, a large scale study was established in 1998. This study, described in **chapter 2**, was performed in a random sample of 1598 adults from a base population of 9012 adult ID service users. This base population was representative for the total population of adult ID service users in the Netherlands. Internationally accepted diagnostic methods and criteria were used. After a re-weighting procedure for non-attenders and a stratification that was applied, a prevalence of hearing impairment of 30% could be estimated for the total population of adult ID-service users. This high prevalence was not only accounted for by the frequently occurring hearing impairment in persons with Down syndrome (prevalence 57%), but also by a prevalence of 24% in the subpopulation with an ID by a different cause, which appeared to be significantly higher than the prevalence of 16-17% in the general population. Apart from the raised prevalence, in the latter subpopulation it also appeared that the prevalence-versus-age curve was shifted one decade forwards to a younger age. Because of this, we want to advise to adjust existing guidelines for hearing screening in adults with an ID by a different cause than Down syndrome: hearing screening on presbyacusis should take place from the age of 40 years instead of 50 years.

As was expected, Down syndrome and age 50 years and over could be identified as risk factors. However, a more serious degree of ID, which is the most important risk factor in visual impairment, could only be identified as a risk factor in the subpopulation with Down syndrome.

Because of the very high prevalence of hearing impairment in adults with Down syndrome, rising to a 100 percent in persons older than 60 years, we want to propose a second adjustment in the aforementioned guidelines: audiometry by an ENT specialist or audiologist should take place every three years instead of hearing screening.

Because the results of an epidemiological study on visual impairment in the same study population already had been presented earlier, we were now able to estimate the nationwide prevalence and severity of dual sensory impairment as well

(**chapter 3**). In 77 participants of the study dual sensory impairment could be diagnosed. After re-weighting the data, a prevalence of 5% could be estimated for the total population of adult ID-service users, of 3% for the age group younger than 50 years, and of 11% for the subpopulation aged 50 years and over. The following risk factors could be identified: a more serious degree of ID, age 50 years and over, and Down syndrome. In three cases the combination of a severe/profound hearing impairment was present in combination with blindness. Treatment or rehabilitation in principle was possible in the majority of cases with dual sensory impairment.

In **Chapter 4**, outcomes are presented of a pilot study into expectations of, and satisfaction with hearing aids in a small groups of adults with a mild or moderate intellectual disability. We wanted to know whether the outcomes were comparable to outcomes of similar studies in the general population. If not, this might have consequences for the introduction and follow-up of hearing aids in this group. Sixteen participants with a recent diagnosis of hearing impairment were interviewed before the introduction of hearing aids and six months afterwards.
It appeared that in a majority, expectations were based on incomplete or incorrect remembered information. Special wishes, e.g. regarding visibility of the hearing aids, influenced acceptation and appreciation in a few cases. Satisfaction with the hearing aids appeared to be related to similar aspects as in the general population: benefit, cosmetics and self-image, sound quality/acoustics, comfort and ease of use, and service (by the hearing aid supplier). Only costs were not an issue here. It was remarkable that some participants explicitly preferred invisible (in-the-ear) hearing aids, whereas others opted for brightly coloured ones. Most of the participants, including those with a mild intellectual disability, appeared to be partially or completely dependent on their caregivers for the use and maintenance of the hearing aids, as well as for detection and solving of problems. In spite of the fact that these caregivers had been specifically trained in this, six months after the introduction of hearing aids, some participants still complained of discomfort, pain and loud noises. As a result, hearing aids were taken out or switched off.
We conclude that: 1. information on hearing loss and hearing aids has to be repeated and checked, 2. specific wishes concerning hearing aids, including cosmetic aspects, should be explicitly asked for, 3. a hearing aids expert (speech and hearing therapist, hearing aid supplier) should be easily accessible and perform regular check-ups to inspect the hearing aids and detect complaints.

Our practical experience was, that treatment with hearing aids in adults with intellectual disabilities quite often fails. It is often assumed that this is caused by client characteristics, such as misunderstanding or fear. However, their environment (dependence on caregivers, unfavourable acoustic circumstances) might also influence acceptation of hearing aids. In **Chapter 5**, we describe a qualitative study into barriers, which were met during the implementation of an adapted hearing rehabilitation protocol by five providers of intellectual disability services (central residential and community-based homes). This adapted rehabilitation protocol had been developed by a Dutch consensus working party. It consisted of four modules: 1. hearing aid fitting and training, 2. optimisation of acoustic circumstances, 3. assessment of communicative skills and needs, followed by multidisciplinary decisions on aspects of the rehabilitation, 4. continuous education of carers (train-the-trainers model). An inventory of problems, expected for the implementation process, was then made by the research team, and strategies were designed to overcome these and discussed with senior management of the service- providers.

In spite of these preparations, within a period of two years all conditions for an optimal audiological rehabilitation could be realised in 3 out 31 participants only. We had expected the implementation problems to occur at the level of clients and direct caregivers, but instead, we already met them at an organisational level, before the hearing aids could be actually fitted. During the implementation process it became clear that senior management (sometimes) had facilitated the interventions, but had taken no measures to ensure continuity. We concluded that we had been able to register the barriers that were met, but did not know what had caused them. In order to study this, an additional study was designed to evaluate the implementation process.

In this second part of the implementation study, qualitative research methods were applied to identify retrospectively the factors that had influenced the implementation process. Twenty-four managers and other professionals from the five participating intellectual disability service-providers and three audiologists were interviewed. Results of this study are presented in **Chapter 6**.

The audiological centres had struggled with a big increase of persons with an ID. In addition they had little experience with performing diagnostics on location of the ID centres. This had caused severe delays. Another problem was that assessment of acoustical conditions in the ID centres had to be performed without financial compensation. Also not all audiologist considered the assessment of acoustics to be their task. The activities of the ID centres and the audiological centres were coordinated insufficiently, and tasks and responsibilities not clearly defined.

The following factors were mentioned by the interviewees from the ID centres: insufficient support by carers and middle management, insufficient documentation of financial and organisational consequences of the project, as well as of local procedures, competition with other projects in the ID centre, insufficient transfer of information, and insufficient embedding in existing local procedures. The quality of transfer of information was reported to be of insufficient quality in the residential as well as in the community based ID centres. The local project coordinators, all professionals, were insufficiently equipped and were insufficiently supported by management to solve these problems.

Organisational culture, reflected in the behaviour of staff and management, may have played a role too, but this is tentative because of the paucity of information. Some of the following themes were identified: focus on quality improvement, focus on results, commitment and responsibility, formal relations between professional disciplines, and the policy of normal versus specialised health care for their clients.

This last theme was mentioned by interviewees from all five participating ID centres. The other themes were mentioned less frequently, but are known from reported study results in other forms of health care, for instance concerning continuous quality improvement in hospitals.

We conclude that audiological rehabilitation in adult users of ID service-providers is a complex intervention with several innovative parts. For a successful implementation, a plan of approach is necessary in which a continual active involvement of senior management is necessary. Expertise in change management is indispensable

We recommend: 1. structured collaboration of audiological centres and intellectual disability service providers, 2. provision of hearing aid habituation and training by audiological centres, and 3. systematic inclusion of continuous education on hearing impairment in education of staff members, and 4. obligatory building standards for acoustical conditions in special schools, homes and day activity centres for persons with an ID, as is already advised by the Netherlands Board for Hospital Facilities. Furthermore we advise management of ID centres to establish projects to improve the quality of care for, and rehabilitation in persons with hearing impairment.

In **Chapter 7**, most relevant findings are presented and commented upon. Furthermore, problems encountered during the studies are discussed: the size of the problem of hearing impairment and of combined hearing and visual impairments in the adult population with an ID, protocols for screening and diagnosis, dependence on carers, and audiological care. A working arrangement is proposed for professionals involved in detection, diagnosis, and rehabilitation of hearing impairment.

Some of the studies in this thesis have a pioneer character, preparing the grounds for in-depth research. We have formulated recommendations for future research, for adaptations in the international IASSID consensus guidelines for early detection of hearing impairments in people with intellectual disabilities, and for structural policies to improve identification and rehabilitation of hearing impairment in intellectual disability centres.

In our opinion, this will lead to an improvement of the quality of audiological care for adults with an intellectual disability.

SAMENVATTING

Inleiding

In dit proefschrift worden de resultaten gepresenteerd van een epidemiologische studie betreffende slechthorendheid bij volwassenen met een verstandelijke beperking, evenals de resultaten van een studie waarin de implementatie van een aangepast behandelingsprotocol voor slechthorendheid in zorginstellingen voor mensen met een verstandelijke beperking werd onderzocht. Het wordt voorafgegaan in **hoofdstuk 1** door een overzicht van de reeds gepubliceerde literatuur op het gebied van slechthorendheid bij mensen met een verstandelijke beperking, zowel wat betreft de epidemiologie als de behandeling. Ook worden in dit hoofdstuk de redenen aangegeven waarom de studies werden opgezet. Daarnaast worden de doelstellingen van de studies geformuleerd en definities gegeven voor verstandelijke beperking, slechthorendheid, slechtziendheid, dubbele zintuiglijke beperking, en implementatie.

Het geringe aantal studies naar prevalentie van slechthorendheid bij volwassenen met een verstandelijke beperking is tot nu toe uitgevoerd in kleine of geselecteerde populaties. Daarnaast werden ook verschillende definities van slechthorendheid gehanteerd. Om de landelijke prevalentie en ernst van slechthorendheid te kunnen bepalen, en risicogroepen te kunnen identificeren, werd in 1998 een grootschalig onderzoek opgezet. Dit onderzoek, dat beschreven staat in **hoofdstuk 2** werd uitgevoerd in een steekproef van 1598 volwassenen uit een basispopulatie van 9012 volwassen gebruikers van woonvoorzieningen en dagcentra voor mensen met een verstandelijke beperking. De basispopulatie vormde een representatieve afspiegeling van de totale populatie van mensen met een verstandelijke beperking in Nederland. Gebruik werd gemaakt van internationaal erkende diagnostische methoden en criteria. Na een re-weighting procedure vanwege uitval tijdens de studie en een toegepaste stratificatie, werd voor de totale populatie van verstandelijk beperkte zorggebruikers in Nederland een prevalentie van slechthorendheid vastgesteld van 30%. Dit hoge percentage bleek niet alleen te worden veroorzaakt door de vaak voorkomende slechthorendheid bij mensen met het syndroom van Down (prevalentie 57%), maar ook door een prevalentie van 24% in de subpopulatie met een verstandelijke beperking door een andere oorzaak, dat nog steeds significant hoger bleek te zijn dan de prevalentie van 16 - 17% in de algemene populatie. Daarnaast bleek dat de prevalentie-versus-leeftijdscurve ongeveer 10 jaar opgeschoven is naar een jongere leeftijd. We adviseren dan ook om in de bestaande Nederlandse en internationale richtlijnen voor gehoorscreening bij volwassenen met een verstandelijke beperking als gevolg van een andere oorzaak dan het syndroom van Down, de screening op ouderdoms-slechthorendheid te laten plaatsvinden vanaf de leeftijd van 40 jaar in plaats van vanaf 50 jaar.

Zoals te verwachten was, vormde, naast Down syndroom, ook een leeftijd van 50 jaar en ouder een verhoogd risico. In tegenstelling tot de situatie bij slechtziendheid, waar een ernstiger mate van verstandelijke beperking de belangrijkste risicofactor is, bleek dit voor slechthorendheid uitsluitend in de subpopulatie met het syndroom van Down een risicofactor te zijn.

Vanwege de zeer hoge prevalentie van slechthorendheid bij volwassenen met het syndroom van Down (ruim 57%), oplopend tot 100% bij 60-plussers, adviseren we een tweede aanpassing in de voornoemde richtlijnen en wel dat elke drie jaar meteen audiometrie wordt verricht door een KNO-arts of audioloog in plaats van dit vooraf te laten gaan door een screeningsronde.

Omdat al eerder de resultaten waren gepresenteerd van een epidemiologisch onderzoek naar slechtziendheid in dezelfde studiepopulatie, konden nu ook de landelijke prevalentie en ernst van een dubbele zintuiglijke beperking worden vastgesteld (**hoofdstuk 3**). Bij 77 deelnemers aan de studie bleek sprake te zijn van een dubbele zintuiglijke beperking. Na re-weighting van de data werd een prevalentie van 5% vastgesteld voor de totale populatie, van 3% voor de leeftijdsgroep jonger dan 50 jaar, en van 11% voor de populatie van 50 jaar en ouder. Deze prevalenties zijn aanzienlijk hoger dan de prevalenties in de algemene populatie. Als risicofactoren konden worden geïdentificeerd: een ernstiger mate van verstandelijke beperking, leeftijd van 50 jaar en ouder, en het syndroom van Down. In drie gevallen was sprake van de combinatie (zeer) ernstige slechthorendheid/ blindheid. De slechthorendheid en slechtziendheid was in principe behandelbaar of revalideerbaar in een meerderheid van de gevallen.

In **hoofdstuk 4** worden de resultaten gepresenteerd van een pilot study waarin verwachtingen van, en tevredenheid met hoortoestellen zijn onderzocht in een groep volwassenen met een verstandelijke beperking. We wilden weten of de resultaten overeenkwamen met resultaten uit vergelijkbare studies in de algemene populatie. Zo niet, dan zou dit consequenties kunnen hebben voor de introductie, maar ook voor de follow-up van de hoortoestellen. Zestien volwassenen met een lichte tot matige verstandelijke beperking werden geïnterviewd vóór het starten met hoortoestellen en een half jaar erna. De verwachtingen bij de cliënten bleken in de meeste gevallen te berusten op incomplete of onjuiste informatie. Speciale wensen ten aanzien van de hoortoestellen, bijvoorbeeld ten aanzien van de zichtbaarheid, bleken in enkele gevallen van invloed te zijn op het uiteindelijke succes van de aanpassing. Tevredenheid met de hoortoestellen bleek samen te hangen met dezelfde aspecten als in de algemene bevolking: opbrengst van de hoortoestellen, cosmetische aspecten en zelfbeeld, kwaliteit van het geluid, comfort en gebruiksgemak, en service van de hoortoestellenleverancier. Opvallend was dat bij sommige deelnemers aan de studie een voorkeur bestond voor zo onzichtbaar mogelijke hoortoestellen, maar bij anderen juist voor helder gekleurde hoorapparatuur. De meeste deelnemers, inclusief enkele mensen met een lichte verstandelijke beperking, bleken gedeeltelijk of geheel afhankelijk te zijn van hun begeleiders in het gebruik en onderhoud van de hoortoestellen. Ondanks het feit dat de begeleiders een training hadden gevolgd om hierin deskundigheid te verkrijgen, inclusief in het signaleren van problemen, bleken bij een aantal cliënten zes maanden na het starten met de hoortoestellen nog steeds klachten te bestaan van ongemak, pijn en geluid. Als gevolg hiervan werden hoortoestellen weer uit het oor gehaald of uitgezet.

Onze conclusies zijn dat 1. informatie meerdere keren gegeven moet worden en dat gecheckt moet worden of het begrepen is, 2. de cliënt gevraagd dient te worden welke specifieke wensen er bestaan ten aanzien van de hoortoestellen, ook ten aanzien van de cosmetische aspecten, en 3. dat een professional (logopedist of audicien) regelmatig bij de cliënten langs dient te gaan om klachten te kunnen signaleren en de hoortoestellen te controleren.

Uit onze ervaring was bekend dat behandeling met hoortoestellen bij volwassenen met een verstandelijke beperking vaak mislukt. Dit kan te maken hebben met cliëntgerelateerde factoren zoals onbegrip of angst, maar ook met factoren die te maken hebben met de omgeving (afhankelijkheid van begeleiders, akoestiek). We wilden weten of het mogelijk was om een adequate behandeling te realiseren in zorginstellingen voor mensen met een verstandelijke beperking. In **hoofdstuk 5** wordt beschreven welke barrières ontmoet werden tijdens het implementeren van een aangepast behandelingsprotocol voor slechthorendheid in vijf zorginstellingen (centrale locaties en gezinsvervangende tehuizen).

Door een werkgroep was allereerst een behandelingsprotocol ontwikkeld, aangepast aan mensen met een verstandelijke beperking, bestaande uit vier modules:
1. hoortoestelaanpassing en een gewennings- en hoortraining, 2. beoordeling van de akoestiek in woon- en werkvertrekken, indien nodig gevolgd door het verrichten van aanpassingen, 3. beoordeling van de communicatieve behoeften en vaardigheden, gevolgd door een multidisciplinaire bespreking in de zorginstelling, en 4. training van groepsleiding volgens een train-de-trainers model. Hierna volgde een inventarisatie van problemen die verwacht werden tijdens het implementatieproces, en werden strategieën ontwikkeld om deze problemen te verhelpen.

Desondanks lukte het in twee jaar tijd maar bij drie van de 31 geïncludeerde deelnemers om alle voorwaarden voor een optimale start met de hoortoestellen te realiseren (gewennings- en hoortraining beschikbaar, groepsleiding getraind, communicatie adviezen besproken, akoestiek verbeterd). We hadden eigenlijk verwacht vooral problemen te ontmoeten op het niveau van de cliënt en begeleiders. In plaats daarvan kwamen we ze al op organisatorisch niveau tegen, voordat daadwerkelijk met de hoortoestellen gestart kon worden. Gaandeweg het implementatieproces werd verder duidelijk, dat het management het (soms) wel mogelijk had gemaakt om tijdens de studie interventies aan te bieden, maar dat geen maatregelen genomen waren om continuïteit na afloop van de studie te garanderen. We concludeerden dat we wel barrières hadden kunnen registreren, maar niet waardoor ze waren veroorzaakt. Om dit te kunnen bestuderen werd een vervolgonderzoek opgezet om het implementatieproces te evalueren.

In het tweede deel van het implementatieonderzoek werden kwalitatieve onderzoeksmethodes gebruikt om retrospectief factoren te identificeren die het implementatieproces hadden beïnvloed. Vierentwintig betrokken medewerkers uit de zorginstellingen (management en andere professionals) en drie audiologen werden geïnterviewd. De resultaten van dit onderzoek staan beschreven in **hoofdstuk 6**.

De audiologische centra hadden te maken met een onverwacht grote aanvoer van nieuwe cliënten. Daarnaast hadden ze vrijwel geen ervaring in het uitvoeren van de diagnostiek op locatie van de zorginstellingen. Dit heeft geleid tot enorme vertragingen. Een ander probleem was, dat het ter plaatse beoordelen van de akoestiek moest gebeuren zonder extra financiële vergoeding, ook vonden niet alle audiologen het hun taak om dit te doen. De activiteiten tussen de zorgorganisaties voor mensen met een verstandelijke beperking en de audiologische centra waren onvoldoende gecoördineerd, taken en verantwoordelijkheden waren niet duidelijk afgesproken.

Voor de zorgorganisaties zelf werden door de geïnterviewden de volgende factoren genoemd, die de implementatie negatief hadden beïnvloed: onvoldoende draagvlak bij groepsleiding en middenmanagement, financiële en organisatorische consequenties van het project onvoldoende in kaart gebracht, lokale procedures onvoldoende in kaart gebracht, competitie met andere lopende projecten, onvoldoende informatieoverdracht, en onvoldoende verankering van het protocol. De kwaliteit van de informatieoverdracht werd zowel op de centrale locaties als in de kleinschaliger woonvormen slecht genoemd. De lokale projectcoördinatoren, allen professionele medewerkers, waren onvoldoende toegerust, en werden onvoldoende door het management ondersteund om deze problemen op te lossen.

Sommige opmerkingen suggereerden, dat het implementatieproces ook beïnvloed zou kunnen zijn door de organisatiecultuur, zoals die naar voren kwam in het gedrag van zowel medewerkers als het management. Op dit vlak werden voorzichtig de volgende relevante aspecten geïdentificeerd: focus op kwaliteitsverbetering, focus op resultaten, betrokkenheid en verantwoordelijkheid van medewerkers, meer of minder formele relaties tussen de disciplines, en het beleid ten aanzien van normale of juist gespecialiseerde gezondheidszorg voor de cliënten. Het laatste aspect werd ter sprake gebracht in alle vijf zorgorganisaties. De andere aspecten werden minder vaak genoemd, maar zijn al langere tijd bekend uit de literatuur over kwaliteitsverbetering in andere vormen van gezondheidszorg, zoals continue kwaliteitsverbetering in ziekenhuizen.

Wij concluderen, dat audiologische revalidatie in deze bevolkingsgroep een complexe interventie is met meerdere innovatieve aspecten. Voor de implementatie is een samenhangend implementatieplan nodig, dat tot stand komt in voortdurend overleg tussen betrokken professionals en het topmanagement. Deskundigheid in verandermanagement is daarbij onontbeerlijk.

Wij adviseren dat 1. zorginstellingen en audiologische centra op gestructureerde wijze met elkaar samen gaan werken, 2. hoortoestelgewennings- en hoortraining vanuit het audiologisch centrum aangeboden gaat worden, 3. deskundigheidsbevordering op het gebied van slechthorendheid structureel opgenomen dient te worden in de basisopleidingen van alle professionals die betrokken zijn bij mensen met een verstandelijke beperking en 4. er bindende bouwvoorschriften komen voor akoestische omstandigheden in scholen, instituten, woningen en dagcentra voor mensen met een verstandelijke beperking, zoals al is geadviseerd door het Bouwcollege. Verder adviseren we het management van zorginstellingen om verbeterprojecten op te zetten op het gebied van de bejegening en behandeling van hun slechthorende cliënten.

In **hoofdstuk 7** worden de belangrijkste bevindingen nog een keer gepresenteerd en becommentarieerd. Verder gaan we in op een aantal problemen die ontmoet werden tijdens de studies. Achtereenvolgens komen aan de orde: de omvang van het gehoorprobleem en van het probleem van de dubbele zintuiglijke beperking, het screeningsprotocol, het diagnostisch protocol, de afhankelijkheid van begeleiders en de audiologische zorg. Een voorstel wordt gedaan voor een samenwerkingsvorm voor professionals die betrokken zijn bij de opsporing en behandeling van slechthorendheid

In dit proefschrift worden enkele studies gepresenteerd met een pionierskarakter. Hiermee is slechts de basis gelegd. In de discussie worden enkele aanbevelingen gedaan voor verder onderzoek. Vervolgens worden de aanbevelingen samengevat enerzijds voor aanpassingen in bestaande richtlijnen voor vroegtijdige opsporing van slechthorendheid bij mensen met een verstandelijke beperking, en anderzijds voor een structurele aanpak van de diagnostiek en behandeling van slechthorendheid in zorginstellingen voor mensen met een verstandelijke beperking.

Wij menen dat hiermee de kwaliteit van de audiologische zorg voor mensen met een verstandelijke beperking zal verbeteren.

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Tegen het eind van 'de rit' werd ik op de been gehouden door de ondersteuningslobby van mijn zussen Marjolein en Jolanda die mij gedurende enkele maanden wekelijks een cadeautje stuurden dat varieerde van een kitscherige afbeelding van Maria van wie het gezicht verdacht veel op dat van een van mijn zussen leek, tot aan een proefschrift met daarin hun lievelingsrecepten.

Van harte wil ik mijn ouders danken die altijd klaar stonden om mij te helpen. Mijn vader is helaas niet meer in staat om de promotieplechtigheid mee te maken, hij overleed eind 2000.

De laatste jaren waren bijzonder zwaar. Lieve Paul, Christiaan en Bart, zonder jullie had ik dit niet volgehouden. Jullie steunden me door dik en dun. Niets was weldadiger dan elke dag weer bij jullie thuis te komen. Ik verheug me op een nabije toekomst waarin weer veel ruimte is voor het 'goede leven' met elkaar, en met familie en vrienden.

OVER DE AUTEUR

Anneke Meuwese-Jongejeugd werd op 25 februari 1955 geboren in Velsen. Zij behaalde in 1972 het diploma HBS-B aan het Ichthus College te IJmuiden. Na een jaar gewerkt te hebben als assistent-groepsleider in een tehuis voor kinderen met een meervoudige handicap ging zij geneeskunde studeren aan de Universiteit van Amsterdam en slaagde in 1980 voor haar artsexamen. In 1981 was zij een half jaar werkzaam als Senior House Officer in het Royal Hospital for Sick Children in Glasgow. Vervolgens specialiseerde ze zich tot kinderarts in het Academisch Medisch Centrum te Amsterdam. Tijdens deze specialisatieperiode was zij ook een half jaar werkzaam op de afdeling kinderoncologie van het Emma Kinderziekenhuis. Na de geboorte van een tweeling werkte ze enkele jaren als schoolarts en was ze als docent verbonden aan de A-opleiding verpleegkunde van het Academisch Ziekenhuis te Utrecht. In de periode van 1990 - 1994 oefende ze het kinderartsenvak uit in de W. Zeylmans van Emmichovenkliniek in Bilthoven. Na het sluiten van het ziekenhuis aanvaardde ze een baan waardoor ze opnieuw ging werken in een zorginstelling voor kinderen met een verstandelijke beperking, maar nu als kinderarts. Vanuit een behoefte aan nieuwe uitdagingen stapte ze na vijf jaar over op de onderzoeksbaan bij de vakgroep geneeskunde voor verstandelijk gehandicapten in het Erasmus Medisch Centrum Rotterdam, in samenwerking met het audiologisch centrum van hetzelfde ziekenhuis, waarvan de werkzaamheden uiteindelijk tot dit proefschrift hebben geleid.

Sinds januari 2005 is Anneke hoofd van de afdeling Vroegtijdige Onderkenning van kinderen met Gehoorstoornissen, bij de Nederlandse Stichting voor het Dove en Slechthorende Kind te Amsterdam.