

**The impact of visual impairment  
on adults with an intellectual disability**



# The impact of visual impairment on adults with an intellectual disability

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to my parents

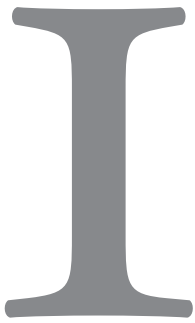




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## **General introduction**



Over forty years ago, the Danish ophthalmologist Mette Warburg was the first to ask attention for the increased risk of visual impairment in people with intellectual disabilities (Warburg, 1963, 1975). She based this on her clinical experience and first evaluations in groups of adults in Danish day care centers. She claimed in an early stage that the risk increased with more severe levels of intellectual disability (Warburg, 1983). Warburg has long remained a voice in the wilderness. Incidental colleagues, among them Lena Jacobson in Sweden, performed ophthalmological evaluations in larger groups and reported an overrepresentation of ocular pathologies (Jacobson, 1988).

However, problems with assessment of visual function in people who were not able to cooperate with Snellen or picture card acuity tests, remained a barrier to identify those with impairments. Gradually, acuity tests using matching cards, developed for assessment in young children by Sheridan & Gardiner (1970) and by Lea Hyvärinen (Hyvärinen et al., 1980), started to be applied in children and adults with moderate intellectual disabilities. In the United States, Davida Teller developed the preferential looking card for assessment of visual acuity in babies and toddlers (Dobson et al., 1978; Teller, 1979), after which Jackie van Hof-van Duin successfully applied this card in Dutch children and adults with severe or profound intellectual disabilities (Mohn & Hof-van Duin, 1983).

The next academic group systematically addressing visual functioning in this population, specifically people with Down syndrome, was the optometry department of Cardiff University in Wales, led by Margareth Woodhouse (Woodhouse et al., 1993). Next to the Teller card, this group developed the Cardiff card, with line pictures in stead of stripes (Woodhouse et al., 1992; Adoh et al., 1992). In this way, visual acuity assessment became feasible in principally all persons with intellectual disabilities who are not able to cooperate with routine diagnostics.

During the 1990's, first consensus guidelines were developed for detection and diagnosis of visual impairment in children and adults with intellectual disabilities in both the Scandinavian countries and the Netherlands (Warburg & Riise, 1994; Nagtzaam & Evenhuis, 1999). This was initiated in the Netherlands by the intellectual disability physician Heleen Evenhuis. In 1996, during a world congress of the International Association on the Scientific Studies of Intellectual Disabilities (IASSID) in Helsinki, a new Special Interest Research Group on Health Issues was founded. One of the first actions of this group was to develop a consensus statement 'Early identification of hearing and visual impairment

in children and adults with intellectual disabilities' (Evenhuis & Nagtzaam, 1998). Active screening was at the heart of these different consensus statements. However, due to the paucity of available research findings, they were only partially based on scientific evidence, and further on joint expert experience.

In the Netherlands, based on the diagnostic criteria and protocol, recommended by the Dutch consensus group, large-scale screening was started by low vision organizations through service providers for people with intellectual disabilities. Meanwhile, Evenhuis prepared, together with the ophthalmologist prof. dr. Jan Stilma (University Medical Center Utrecht), a large multi-center epidemiologic study, to quantify the risk and identify specific risk groups. During 1999 – 2003, around 1600 adults with intellectual disabilities in care organizations all over the country underwent on-site diagnostic assessments by PhD students, together with diagnostic experts of Bartiméus, a large Dutch organization for people with low vision and blindness. The results were valid and striking, showing that the risk of visual impairment as well as of blindness in adults aged 18 years and over was ten times increased, as compared to the risk in older adults aged 55 years and over in the general Dutch population (Splunder et al., 2006; Klaver et al., 1999). The diagnosis had not been detected prior to the study in 43% of participants with visual impairment and in 36% of participants with blindness. All subgroups had an increased risk, but people with severe intellectual disabilities and, to a lesser extent, people with Down syndrome and of course older people were specifically affected. Many untreated but treatable causes, such as refractive errors (need of spectacles) and cataract, were identified (Splunder et al., 2004). The shameful conclusion must be that even in high-income countries, avoidable visual impairment and blindness are highly prevalent among people with intellectual disabilities.

In the meantime, Snowdon and Stewart-Brown (Snowdon and Stewart-Brown, 1997; Stewart-Brown & Snowdon, 1998) concluded on the basis of a systematic literature review, that the ethical basis for preschool vision screening in the general population was very weak, 'in the absence of sound evidence that the target conditions sought in these programs are disabling and that available interventions to correct them do more good than harm'. Does this also apply to screening in the population with intellectual disabilities? The risk has now been validly quantified for adults (Splunder et al., 2006) and risk groups have been identified. Treatable ophthalmological conditions can be diagnosed in many cases, of which refractive errors and cataracts are the most prevalent (Splunder et al., 2004). Reliable measurements of visual acuity, refraction and slit lamp

biomicroscopy appear feasible in a large majority of adults with mild to profound intellectual disabilities (Splunder et al., 2004). With this scientific evidence, part of Wilson's criteria for population screening (Wilson & Jungner, 1968) have been met, but not all. How disabling is visual impairment in this group, on top of the intellectual disability? Do people with intellectual disability prefer and accept treatment of visual impairment with glasses or surgery? Which is the quality of diagnostic and therapeutic follow-up? It can be claimed that, at least in high-income countries, facilities for assessment and effective treatment are sufficiently available. However, it is unclear whether in practice these facilities are really available to persons with intellectual disabilities. Is there effective diagnostic follow-up after screening? Is ophthalmological treatment, or in case of untreatable visual impairment, rehabilitation adequately executed? How effective and accepted are these interventions? For which groups? Is there really a case for screening in this population, or in subgroups?

Therefore, a logical next step was to study these questions. This resulted in the current PhD project, started in 2001, with the primary aim to study effects of low vision as well as effects of treatment and rehabilitation of visual impairment in adults with intellectual disabilities.

We decided to design a randomized controlled trial, as this is the most valid design to study treatment effects. A very large base population was required to obtain a study sample with low vision that would be large enough for randomization. We have approached 15 care providers for people with intellectual disabilities, of whom three consented to participate. Although medical treatments would be reimbursed by health insurances, this was not the case with rehabilitation costs (optimizations of illumination and contrasts, time of caring staff for specific support) and costs of replacement during training of staff. The fact that our study budget did not permit to finance such costs, as well as the fact that time of carers would be required to collect observational information on outcome measures, have certainly contributed to the low enthusiasm to participate. However, the three care providers that were prepared to support the study together had around 5000 clients, which with a prevalence of 20% low vision and blindness seemed a large enough base population. Low vision teams of Sensis, De Brink and Bartiméus were prepared to cooperate and if necessary modify their routine screening schedules and diagnostic protocols.

It is regularly to be heard, that general practitioners and ophthalmologists, but also carers, doubt whether in this already functionally disabled population

low vision will lead to extra disability. With baseline measures of functional and behavioral aspects in 269 eligible adult clients with and without low vision, we were able to for the first time address this topic (Chapter 2).

In Chapter 3, results are presented of a systematic literature review into feasibility and functional effects of treatment of refractive errors, cataract and other ophthalmological conditions, as well as rehabilitation of low vision and blindness, in children and adults with intellectual disabilities or related conditions (cerebral palsy). It appeared that the existing scientific knowledge is poor in the population with ID but also in the general population evidence for usual treatments is not yet established.

Interventions were heterogeneous, and could consist of medical treatment (spectacles, cataract surgery), technical optimization of illumination, contrasts and routing, and expert support by specifically trained carer teams. To obtain sound evidence, it was important that advised interventions were realized. Prior to the start of diagnostic assessments and data collection, we invested time and energy to support and stimulate implementation by staff in residential homes, with organizational and financial support by the management. How we did this, will be described in Chapter 4, whereas in Chapter 5, the actual implementation in the intervention group and factors influencing implementation will be presented.

Recently, a retrospective Dutch study has been published, based on files of Bartiméus during over ten years (Isterdael et al., 2008). Treatment advice for refractive errors and cataract and their implementation in 6220 referred clients with intellectual disabilities were presented. Chapter 6 is a comment on this study.

Chapter 7 finally addresses the primary study question of the trial: effects of treatment and rehabilitation.

The General Discussion in Chapter 8 will be used to give overall comments on the findings of this trial and to describe side effects of the study on ideas, procedures and priorities of Dutch low vision teams for people with intellectual disabilities.







# 2

**Does visual impairment lead to additional disability in adults with intellectual disabilities?**

**A cross-sectional study**

Sjoukes L, Koot JM, Kooijman AC & Evenhuis HM

## Abstract

**Background** This study addresses the question to what extent visual impairment leads to additional disability in adults with intellectual disabilities (ID).

**Method** In a multi-center cross-sectional study of 269 adults with mild to profound ID, social and behavioral functioning was assessed with observant-based questionnaires, prior to expert assessment of visual function. With linear regression analysis the percentage of variance, explained by levels of visual function, was calculated for the total population and per ID level.

**Results** 107/269 participants were visually impaired or blind (WHO criteria). On top of the decrease by ID visual impairment significantly decreased daily living skills, communication & language, recognition and communication. Visual impairment was not correlated with more self-absorbed and withdrawn behavior or anxiety. Peculiar looking habits correlated with visual impairment and not with ID. In the groups with moderate and severe ID this effect seems stronger than in the group with profound ID.

**Conclusion** Although ID alone impairs daily functioning, visual impairment diminishes the daily functioning even more. Timely detection and treatment or rehabilitation of visual impairment may positively influence daily functioning, language development, initiative and persistence, social skills, communication skills, and insecure movement.

## Introduction

When intellectual disability (ID), which in itself seriously afflicts psychosocial development and daily functioning, co-occurs with a visual impairment, it may be expected that overall disability will be more severe. Co-occurrence of intellectual and visual impairment is not uncommon: Splunder et al. (2006) have shown that 14% of the adult population with intellectual disabilities (ID) has a visual impairment and 5% is (socially) blind.

The general effects of ID and visual impairment are well documented. ID impairs activities of daily life (ADL), linguistic skills, social skills and independent living skills (Kottorp et al., 2003). Such impairments remain relatively stable over time (Beadle-Brown et al., 2002), as does stereotyped behavior (Thompson & Reid, 2002). Childhood visual impairment delays motor development (Precht et al., 2001) and complicates and delays attachment, language, motor and learning skills (Levtzion-Korach et al., 2000; Dale & Sonksen, 2002; Atkinson et al., 2002). Stereotyped behavior like body-rocking, repetitive handling of objects, and eye-pressing or eye-poking is common (Dijk 1983; Jan et al., 1983; Fazzi et al., 1999).

Visual impairment in the general adult population interferes negatively with the ability to perform ADL (Haymes et al., 2002). At all ages, sleep problems have been reported for people with (profound) low vision or blindness (Brylewski & Wiggs, 1999; Dale & Sonksen, 2002, Didden et al., 2002; Zizi et al., 2002). Studies in groups aged 65 years and over have shown that the third most common condition causing a need for ADL assistance is age-related vision loss after arthritis and heart disease and this may lead to depression, anxiety, lethargy and social dissatisfaction (Shmueli-Dulitzki & Rovner, 1997; Keller et al., 1999; Heine & Browning, 2002; Warnecke, 2003). However, as far as we know, no research has been published on the extra disability due to visual impairment in the population with ID. On the contrary, visual impairment and even blindness tend to remain 'invisible' in this population: in 40% of Dutch adults with ID the visual impairment (or blindness) had not been identified (Splunder et al., 2006). Possible explanations might be that persons with ID do not complain of visual loss or that symptoms of visual impairment are labelled as originating from the ID or autism. Another possibility is, that those with low vision show less acting-out behavior or ask less attention than people with ID and normal vision, thus suggesting that everything is going well.

This study addresses the question to what extent visual impairment leads to additional disability in adults with ID. We hypothesized that independent of the severity of ID, lower vision is associated with:

- Decreased independent living skills;
- Increased difficulties in communication;
- More insecure movement;
- More self-absorbed and withdrawn behavior;
- Less antisocial and disruptive behavior.

## Methods

### Study design and procedure

A multi-center cross-sectional study design was used to assess the relationship between the participants' levels of ID and visual functioning on the one hand, and behavior and social functioning on the other hand.

The procedure is generally outlined here; details are given below. Both ID and visual function were categorized into four classes (Table 2.1). Behavior and social functioning were assessed with observational questionnaires that addressed the following five areas: independent living skills, communication, insecure movement, peculiar looking habits, self-absorbed behavior, and antisocial, disruptive behavior. Caregivers who knew the participants well completed these questionnaires. This was done prior to the screening of visual function, so caregivers were blind to the actual visual function of the participants. The study was approved by the Medical Ethics Committee of the Erasmus University Medical Center (MEC 197.075/2000/239).

### Study population

Three Dutch ID Service providers offering sheltered workshops, residential and day care, where a routine vision screening by a district low vision center had been scheduled for 2002 – 2004, consented to participate in the study. The three involved low vision teams screened according to consensus guidelines of the International Association on the Scientific Studies of Intellectual Disability (IASSID) (Evenhuis & Nagtzaam, 1998). Because it was important to include enough participants in the different ID and visual function categories, we aimed to include both homes for clients needing care and homes for more independent

clients in the screening. Because of the limited capacity of the low vision teams with their highly specialized staff, caregivers were asked to complete a short, non-validated observation-based questionnaire to estimate the possibility of visual problems. Only clients with a possibly elevated risk of visual problems were invited for the screening. Criteria for inclusion in the study were: age 18 years and over, no previous diagnosis of visual impairment, and no severe motor impairment or progressive disease (e.g. dementia or cancer). Written informed consent for participation was asked from the legal representatives and from clients who were able to understand the aims of the study.

### **General data**

Information on the severity of ID, based on prior IQ testing, was obtained from the patient files, just as information on relevant co-morbidity (disruptive behavior, hearing impairment, etiology of ID, motor impairment and wheelchair use).

### **Visual function assessment**

Visual functioning was assessed on-site in the homes or day activity centers, according to IASSID consensus guidelines (Evenhuis & Nagtzaam, 1998), by professionals of the regional low vision teams with a specific expertise on people with ID. Assessment included visual acuity with at least two tests (Snellen's chart, Burghardt's children's chart, STYCAR single characters and matching, Cardiff Acuity Cards, Teller Acuity Cards), visual fields (confrontation), contrast sensitivity (Hiding Heidi), auto refraction or sciascopy, and assessment of strabismus. Referral for ophthalmological diagnosis and treatment was advised if necessary.

### **Definitions and classifications**

All participants were assigned to one of the ID and vision categories, as presented in Table 2.1. For this study, visual function was defined as the visual acuity in the best eye (with optimal correction, if glasses were used). 'Visual impairment' and '(social) blindness' were classified according to criteria of the World Health Organization (WHO) (1980).

Table 2.1  
Classification of intellectual disability (ID) and visual function

ID level	IQ	Developmental age [years]	Visual function level	Visual acuity [arcmin <sup>-1</sup> ]
Mild	50 – 70	6 – 11	Normal vision	≥ 0.5
Moderate	35 – 50	3.5 – 6	Mild impairment	0.3 – 0.5
Severe	20 – 35	1.5 – 3.5	Visual impairment	0.05 – 0.3
Profound	< 20	< 1.5	(Social) blindness	< 0.05

### Behavioral questionnaires

*Adaptive behavior scale (SRZ).* Adaptive behavior was measured with the Dutch ‘Sociale Redzaamheidsschaal voor Zwakzinnigen’ (SRZ) (Social Functioning Scale for people with ID) (Kraijer & Kema, 1994). The questionnaire consists of 31 items, each with four descriptions of possible behavior, from inapt to adequate, with corresponding rating values from 1 to 4. The item score is the rating value of the description that fits a participant’s behavior best. The SRZ total score is the sum of all ratings and ranges from 31 – 124. Adaptive behavior is further specified with four subscales in which items are grouped: daily living skills (12 items), initiative & persistence (5 items), social skills (5 items) and communication & language development (9 items). The SRZ is found to be reliable and well validated (Kraijer & Kema, 1994). The correlation with the Vineland Social Maturity Scale is 0.82 (Kraijer, 2000). The instrument is widely used in the Netherlands for persons with moderate and severe ID aged 4 years and over.

*Vision Related Behavior Questionnaire (VRBQ).* The Vision Related Behavior Questionnaire (VRBQ) is a new questionnaire for the assessment of specific vision-related behavior in people with ID that we developed because there was no suitable instrument available for the population of this study. An expert group consisting of behavioral scientists from the three participating low vision expertise centers was formed to define examples of vision-related behavior. The questionnaire consists of 45 behavioral items pertaining to problems with eye-hand coordination, eye-foot coordination, recognition of persons and objects, spatial orientation, insecure movement, anxiety, peculiar looking habits and communication. The items are scored in terms of occurrence: always, often,



sometimes or never present, with corresponding rating values 1 to 4. A higher rating corresponds to more normal behavior.

Five clusters of vision-related behavior were identified using principal component analysis (based on 41/45 items with a factor loading  $\geq 0.4$ ): eye-hand coordination & insecure movement (9 items), recognition & communicative skills (9 items), eye-foot coordination (8 items), anxiety/ fear (6 items) and peculiar looking habits (9 items). The maximum score for each subscale is the number of constituent items multiplied by 4. The VRBQ total score is the sum of all individual rating values and ranges from 45 to 180. The VRBQ has been shown to be reliable: Cronbach  $\alpha$  is 0.6383 (a scale is reliable when Cronbach's  $\alpha > 0.6$  (Malhotra, 1993).

*Development Behavior Checklist (DBC).* The Development Behavior Checklist (DBC) was developed to identify emotional and behavioral problems in children with ID (Einfeld & Tonge, 1995). It has been translated and validated for Dutch children, but it proved to be applicable to adults as well (Dekker et al., 2002; Ruiters et al., in press). The DBC includes 95 descriptions of problem behavior that are scored as either not true, partly true, or true, with the rating values 0, 1 and 2. Higher scores indicate more emotional and behavioral problems. The DBC has the following five, partly overlapping subscales: disruptive & anti-social behavior (27 items), self-absorbed behavior (31 items), communication disturbance (13 items), anxiety (9 items), and social relating (10 items).

### Analysis

The relation between visual function and ID levels was analyzed with Spearman's correlation to determine to what extent ID is a predictor of visual impairment. Means and standard deviations of the questionnaire scores were computed for the ID and for the visual function levels separately. Spearman's  $\rho$  was calculated for the questionnaire total and subscale scores with levels of visual function and ID.

We investigated the effect of vision problems with linear regression adjusting for ID level, disruptive behavior, Down syndrome, hearing impairment, wheelchair dependency, and age. We computed the unstandardized  $\beta$  to determine the direction of the effects. To refine these results we repeated the linear regression analysis per ID level. The relevance is described using Cohen's criteria: effect sizes are considered small if the percentage of explained variance (PEV) is

< 5.9%, medium if PEV is between 5.9% and 13.0% and large if PEV exceeds 13.0% (Cohen, 1988).

## Results

### Study population

Informed consent for participation was obtained of 277 clients, 63% female, median age 46 years, range 17 – 79 years. In eight cases, no visual function test was performed because of organizational problems. This left a study population of 269 persons. Etiology of ID was Down syndrome in 45 cases (16.2%), autism spectrum disorders 24 (9.0%) (as mentioned by the psychologist), 25 other diagnoses in 49 cases (18.4%), and unknown in 156 cases (58.0%). The following co-morbidities were diagnosed: hearing impairment 49 (of whom 14 had hearing aids), no hearing impairment 124, hearing function unknown 96; disruptive behavior 77, no disruptive behavior 154, no data on behavior 38, 14 cases were wheelchair dependant (11 in the group with profound ID and 1 in each of the other ID groups)

Levels of ID and visual function are presented in Table 2.2. Of the 269 participants, 107 were visually impaired or blind according to WHO criteria. There were only 11 participants with a mild ID, because most clients of the participating homes had a moderate or severe ID.

Table 2.2  
Distribution of visual function levels over intellectual disability (ID) levels  
(N = 269)

	Mild ID	Moderate ID	Severe ID	Profound ID	Total
Normal vision	9 (82%)	46 (49%)	39 (32%)	6 (14%)	100
Mild impairment	2 (18%)	25 (26%)	23 (19%)	12 (29%)	62
Visual impairment	0 (0%)	21 (22%)	55 (45%)	18 (43%)	94
(Social) blindness	0 (0%)	2 (2%)	5 (4%)	6 (14%)	13
Total	11	94	122	42	269

## Outcomes

Response rates for the SRZ, VRBQ and DBC questionnaires were 86%, 89% and 89%, respectively, but not all questionnaires were filled out completely. There were no significant differences in age or sex between responders and non-responders.

Gender had no significant effect on any outcome and was not taken into account in the further analysis.

The interdependence of ID levels and visual impairment is illustrated in Table 2.2. Regression analysis yielded a Spearman's correlation  $r_s = 0.3435$ , meaning that visual impairment is predicted to an average degree by the level of ID.

Mean scores and standard deviations of all total and subscales of the SRZ, VRBQ and DBC were calculated (data not shown). The differences in mean scores between those with normal vision and those who were (socially) blind, were 1.6 standard deviation (SD) for SRZ total scores, and 1.8 SD for VRBQ total scores, both significant at  $p = 0.000$ , showing that visual impairment lowers the SRZ and VRBQ total scores.

The results of the regression analysis of the total study population are presented in Table 2.3 and shown graphically in Figure 2.1.

Intellectual disability decreases strongly the adaptive behavior as measured with the SRZ. A large effect was found on the VRBQ total and recognition/communicative scales and a moderate effect on eye hand coordination/ insecure movement.

The SRZ total scores show moderate and significant negative correlations with visual function as well as strong correlations with severity of ID, indicating that adaptive behavior decreases with increasing intellectual and visual impairment. Similar correlations were found for all SRZ subscale scores. The VRBQ subscale scores show smaller, but significant correlations with both visual function and severity of ID. This means that vision-related behavior, like eye-hand coordination/ mobility, and recognition/ communicative skills and secure movement become more difficult, whereas peculiar looking habits increase with increasing visual impairment and more severe ID.

Table 2.3  
 Linear regression analysis of total study population  
 with percentage of explained variance (PEV) of vision level, with ID level,  
 disruptive behavior, Down syndrome, mobility<sup>1</sup>, hearing impairment  
 and age as covariates

(sub)scale	Percentage of explained variance by						
	Vision level	ID level	Disruptive behavior	Down syndrome	Wheelchair dependant	Hearing impairment	Age
SRZ total	5.9**	52.1**	1.4*	.	.	.	.
Daily living skills	6.7**	44.6**	.	.	.	.	.
Communication & language	3.9**	47.3**	.	.	.	.	.
Initiative & persistence	3.9**	31.8**	.	.	.	.	.
Social skills	4.1**	37.7**	3.6**	.	.	.	.
VRBQ total	14.9**	29.5**	.	.	.	.	.
Eye.hand coordination/ mobility	9.5**	5.6**	.	.	28.7**	2.7**	2.7**
Recognition/ communicative skills	8.9**	47.5**	2.0**	.	.	.	.
Insecure movement	6.8**	.	.	.	5.0**	.	.
Anxiety & fear	.	.	.	.	.	.	.
Peculiar looking habits	7.6**	3.0*	.	.	.	.	.
DBC total	.	.	29.6**	.	.	.	.
Disruptive & antisocial	.	.	19.5**	2.4*	.	.	.
Self.absorbed behavior	.	.	28.2**	.	.	.	.
Communication disturbances	7.4**	.	4.7**	.	.	.	3.7**
Anxiety	.	.	.	.	.	.	.
Social relating	.	.	15.1**	4.5**	.	.	.

<sup>1</sup> mobility as the presence or absence of wheelchair dependency

\* significance (p) < 0.05

\*\* significance (p) < 0.01

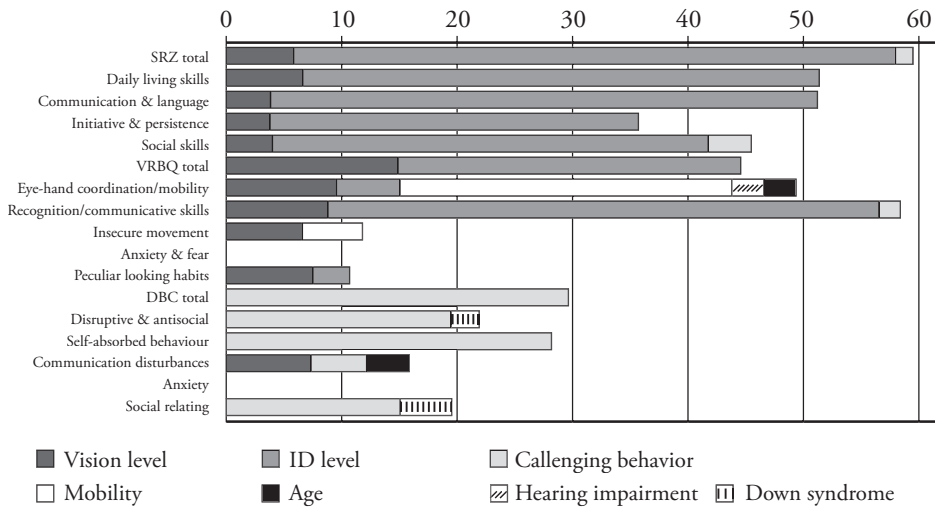


Figure 2.1 Graphical representation of linear regression analysis indicating the relative contribution of covariates.

The diagnosis of disruptive behavior shows strong effects on DBC scores as could be expected, and is related to a small decrease of social skills on the SRZ. Visual impairment does not have relevant effects on the behavioral pathology as measured with the DBC.

People with Down syndrome scored lower on disruptive/ antisocial behavior and better on social relating scales of the DBC.

Hearing impairment and age only had small effects.

It appears that generally, ID has larger behavioral effects than visual impairment. However, there are distinct effects of visual impairment, whereas some subscales are influenced by visual impairment only.

The interdependence of ID level and visual impairment made it necessary to determine the behavioral effects of visual impairment for each ID level separately. Results of the regression analysis per ID group with both vision level and the diagnosis of disruptive behavior as variables are shown in Table 2.4.

Table 2.4

Regression analysis per ID group with percentage of explained variance (PEV) and (the unstandardized  $\beta$ ) of Vision level, with Disruptive behavior as covariate

(sub)scale	ID group					
	Moderate (n = 94)		Severe (n = 122)		Profound (n = 42)	
	Vision level	Disruptive behavior	Vision level	Disruptive behavior	Vision level	Disruptive behavior
SRZ total	22.3** (-12.253)	.	13.3** (-5.252)	.	23.3** (2.702)	.
Daily living skills	12.3** (-4.737)	.	14.4** (-2.820)	.	16.6* (1.411)	.
Comm. & language	12.2** (-3.193)	.	8.5** (-1.593)	.	12.3* (1.048)	.
Initiative & persistence	12.2** (-2,055)	.	.	.	.	.
Social skills	17.0** (-1,735)	.	.	.	.	.
VRBQ total	23.8** (-7.967)	.	12.2** (-4.622)	.	.	.
Eye-hand coord/mobility	7.5** (-1.362)	.	15.2** (-2.081)	.	.	.
Recogn./comm. skills	16.1** (-2.479)	.	20.4** (-2.112)	.	.	.
Insecure movement	.	.	.	.	.	.
Anxiety & fear	.	.	.	5.6* (-1.580)	.	.
Peculiar looking habits	7.0* (-1.016)	.	6.3* (-0.817)	.	.	.
DBC total	.	16.8** (24.400)	10.5* (-7.560)	27.4** (22.439)	.	.
Disruptive & antisocial	.	23.6** (9.881)	3.4* (-1.493)	24.2** (6.535)	.	14.2* (4.886)
Self-absorbed behavior	.	11.2** (7.375)	.	22.6** (9.087)	.	.
Comm. disturbances	.	.	19.3** (-1.542)	.	.	18.3* (2.786)
Anxiety	.	.	5.1* (-0.636)	.	.	.
Social relating	.	17.1** (3.354)	8.1** (-0.987)	.	.	.

\* significance (p) < 0.05

\*\* significance (p) < 0.01

The group with mild ID was excluded because of its small size ( $N = 11$ ), resulting in non-significant effects. Significant effects of visual impairment were found in all other ID level groups, with the largest number of effects in the largest groups (moderate and severe ID) and only on one item in the group with profound ID.

In the group with moderate ID ( $n = 92$ ) all SRZ scores show large percentage of explained variance (PEV), indicating less adaptive behavior with lower vision. The VRBQ total score and the subscale score of recognition/communicative skills were influenced negatively by lower vision whereas peculiar looking habits increased. Lower visual acuity does not show relevant effects on DBC total and sub scores as opposed to a diagnosis of disruptive behavior.

In the group with severe ID ( $n = 122$ ) a similar overall pattern is visible for SRZ and VRBQ scores as in the group with moderate ID, however the effects of the visual function are smaller. Again the DBC-scores are influenced by a diagnosis of disruptive behavior, but not by low vision.

In the group with profound ID ( $n = 42$ ) no effect of vision level could be shown on any scale. The diagnosis of disruptive behavior increased the SRZ total score and subscale scores daily living skills and communication and language, and the DBC scale scores for disruptive behavior (not surprising) and communication disturbances.

## Discussion

This first cross-sectional study, investigating whether visual impairment leads to extra disability for adults with intellectual disability (ID), shows that indeed this is the case. It is true that independent living skills, communication and social skills are primarily determined by the severity of ID. However, visual impairment has an additional influence (Figure 2.1). The presence of disruptive behavior increased the DBC scores strongly, as could be expected. In subgroups with moderate ID in accordance with our first hypothesis visual impairment leads to a large decrease of independent living skills (daily living skills, communication and language, initiative and persistence, social skills). Our second and third hypothesis that communication would become more difficult and insecure movement would increase with lower vision could also be confirmed. Contrary to our fourth

hypothesis, visual impairment is not related to more self-absorbed behavior and in the subgroup with severe ID lower vision is even associated with less anxiety. Our fifth hypothesis that disruptive and antisocial behavior would decrease with lower vision was not confirmed. The presence of disruptive behavior was the most important factor for all DBC outcomes.

In the group with profound ID the above effects were not shown. The group size ( $n = 42$ ) played a role, but another explanation can be that this is the group of people with the most limited daily living skills, therefore the need help with almost everything in daily life. Since the burden of visual impairment is dependent of the requirements of the environment it may be that for people with profound ID low vision is not perceived as a problem, because always carers offer help.

We only included people who had not previously been diagnosed with low vision. This may have influenced our outcomes negatively, because maybe there are differences in behavior between the group with early-diagnosed visual impairment and our research population that led to early diagnosis of visual impairment.

Most findings are basically in concordance with published effects of visual impairment and blindness in the general population, both in children and in ageing people, as has been elucidated in the Introduction. Only the findings of more depression, lethargy and anxiety, observed in persons with age-related visual impairment in the general population (Keller et al., 1999; Heine & Browning, 2002; Warnecke, 2003) could not be confirmed for the adult population with ID. This might be partially explained by the fact, that in the studied population, the newly diagnosed visual impairments may have existed since childhood.

A factor that may have influenced our outcomes was our selection of participants with a possibly increased risk of visual impairment, with a questionnaire for caregivers on observable behavior that supposedly indicates low vision. In fact, to increase effectiveness of large-scale screening of clients with ID, all Dutch low vision centers use such questionnaires. None of these questionnaires has been validated, and indeed, 60% of the participants (162/269) had no visual impairment. Nevertheless, in spite of a selection of the study group towards behavior suspect for low vision, significant differences between groups with and without visual impairment have been demonstrated. We conclude that assessment of a completely unselected population might have resulted in even more significant outcomes.



The present study was a cross-sectional investigation of an adult group with a mix of ages, of severities of ID, and of congenital, childhood and age-related visual impairments, giving first ideas on extra disability as a result of visual impairment. We could not pass a judgement on people with a mild ID. Diagnosed and undiagnosed co-morbidities may have influenced the outcomes: the presence or absence of disruptive behavior appeared influence the outcomes of the DBC much more than low vision does. Down syndrome, hearing impairment and age did not make much difference.

As long as ten years ago, the special interest group 'Health Issues' of the International Association on the Scientific Studies of Intellectual Disability (IASSID) has developed consensus guidelines for the active detection and diagnosis of visual and hearing impairment (Evenhuis & Nagtzaam, 1998). As far as the size of the risk is concerned, this guideline is now sufficiently evidence-based (Splunder et al., 2006). It has led to the organization of large-scale screening in the Netherlands, but nevertheless, around 40% of both visual impairment and blindness remain unidentified (Splunder et al., 2006). The findings of this study explain to some extent why this is the case. People with visual impairment do not show more acting out behavior and are not more anxious than others, so in the perspective of caregivers, they do not cause problems and appear at ease, whereas the fact that they need more help in activities of daily life may not be a problem to professional caregivers. This explains why for the management of service providers, active detection and rehabilitation of visual impairment have a lower priority than the management of motor impairment, epilepsy, autism or disruptive behavior.

Controlled prospective studies in children and in older people would produce more valid insight into the effects of visual impairment. Nevertheless, this study has provided a first scientific basis for the expectation, that in groups especially with mild and moderate ID timely detection and treatment or rehabilitation of visual impairment and blindness may have a positive influence on daily living skills, language development, communication skills, initiative and persistence, social skills, and insecure movement. In this way, it supports evidence for the existing IASSID consensus guideline, stressing the importance of the implementation of its recommendations.



# 3

## **Feasibility and effects of treatment and rehabilitation of low vision in persons with intellectual disabilities**

Sjoukes L & Evenhuis HM



We have shown in previous research, that adults with intellectual disabilities (ID) aged 18 years and over have a ten times increased risk of both visual impairment and blindness (WHO criteria), as compared to the general Dutch population aged 55 years and over (Splunder et al., 2006; Klaver et al., 1998). Until now, this risk has not been quantified in children with ID. It is to be expected that this will be increased as well, because in children's populations with low vision or blindness a majority has an intellectual disability (Mervis et al., 2002).

Causes of low vision in the population with ID often are treatable ophthalmological conditions: severe refractive errors, cataract, strabismus, keratoconus and corneal opacities, glaucoma (Splunder, 2004). Several of these conditions occur specifically in children and adults with Down syndrome, but not exclusively. Further, there is an increased risk of untreatable retinal pathology, optic atrophy and cerebral visual impairment in this population, as well as infrequent other eye pathology. Cerebral visual impairment and retinal pathology are also frequently encountered in persons with cerebral palsy and some of them also have intellectual disabilities.

Life expectancy of people with ID considerably increased during the past decades (Patja et al., 2001) and age related ophthalmological diagnoses can be expected to increase simultaneously. It is well known that in persons with Down syndrome age-related cataract may develop around the age of 30 years (Jaeger, 1980, Splunder et al., 2004). Age-related refractive errors, cataract and glaucoma may result in a relatively early decline of vision: in persons with intellectual disabilities without Down syndrome, clinically relevant cataract becomes apparent 10-15 years earlier than in the general ageing population (Splunder et al., 2004).

We have recently found that, apart from the effects of intellectual disability, visual impairment has significant additional effects on functioning and behavior in adults with moderate, severe or profound intellectual disabilities (Sjoukes, in press). This concerns specifically daily functioning, communication and language, mobility and typical vision-related behavior. It is therefore likely that timely prevention treatment and rehabilitation will lead to improvement of functional limitations and normalization of behavior.

Although many older persons with intellectual disabilities are not able to read, reading glasses may be indispensable for looking at pictures and photos, close up work and day activities, like knitting, embroidering, and jig sawing.

Rehabilitation, specific support by caregivers and an optimal living environment (illumination, contrasts, routing) can be offered in case of untreatable conditions.

A major problem in practice is that general practitioners, ophthalmologists, family and caregivers of people with intellectual disabilities often doubt the feasibility and effects of treatment and rehabilitation in these psychologically vulnerable patients and are reluctant to burden them with it.

In this systematic review we want to offer physicians, ophthalmologists and professional carers information to support the decision process of whether or not to start treatment.

### **Objective**

To evaluate functional effects of treatment and rehabilitation for ophthalmological conditions, oculomotor problems, visual impairment and blindness in children and adults with intellectual disabilities. And consequently compare the results with the treatment in the general population to find arguments pro or against treatment of people with ID in the same way.

## **Criteria for inclusion of studies for this review**

### **Types of studies**

- We searched for intervention studies in populations with intellectual disabilities, and because of overlapping problems also persons with cerebral palsy (Guzetti, 2001) all retrospective and prospective studies and reviews on effects and feasibility of interventions were accepted;
- Cochrane reviews and recent other systematic reviews are studied, to obtain information on effective treatments in the general population. If no systematic reviews were available, other recent literature was evaluated.

### **Types of participants**

Children and/or adults with mild to profound intellectual disabilities or cerebral palsy, with one or more of the following conditions: refractive error, cataract, strabismus, amblyopia, glaucoma, keratoconus, visual impairment or low vision or blindness. Exclusion criteria: none.

The same conditions as above were searched for treatment in the general population.

### **Types of intervention**

All therapies and types of rehabilitation for the following conditions: refractive errors, cataract, treatment of strabismus, glaucoma, technical adaptations in the living environment, all instructions and training programs for patients and/or caregivers.

### **Outcome measures**

- Effectiveness: proportions of participants who did improve at follow-up in visual acuity, functional measures, behavioral measures;
- Feasibility: proportions of participants in which treatment or rehabilitation was not tried, not consented, not complied with, failed, not completed or discontinued, or participants experiencing adverse effects or complications. Similar outcome measures on feasibility were sought for the general population.

### **Search methods for identification of studies**

For the systematic review in at risk groups, the following search terms were used to identify relevant studies listed on the electronic databases:

- Eye diseases (refractive errors, cataract, strabismus, amblyopia, glaucoma, keratoconus), eyeglasses, low vision, cerebral visual impairment;
- Mental retardation, cerebral palsy, Down syndrome.

Search terms:

- “Eye diseases”[Mesh] AND (“Mental Retardation”[Mesh] OR “Cerebral Palsy”[Mesh]).

For the review in the general population all reviews on interventions of any kind were included. In both populations we did not except pharmaceutical trials nor comparisons of surgical techniques.

We identified studies from the following sources:

- The Cochrane Controlled Trials Register (CENTRAL);
- MEDLINE 1966 to mid 2008;
- The lists of references in relevant publications.

## Methods of the systematic review

### Study selection

From the title and abstract we reviewed literature searches to identify potentially relevant studies for full review. Searches of bibliographies and texts were conducted to identify additional intervention studies. The following information was recorded: study setting, year of study, study design, patient recruitment details, inclusion and exclusion criteria, type of intervention, number of included participants, representativeness of study population, duration of follow-up, outcome measures, aspects of feasibility, methodological quality.

### Quality assessment

The following components of quality were assessed:

- Study design of SIGN (BMJ 2001;323:334–336);
- Grades of recommendations (Spine 2007;32(19S):S66–S72);
- Size, power and representativeness of study population;
- Duration of follow-up;
- Completeness of information on consent, inclusion, participation, reasons for failure;
- Validity of outcome measures;
- Quality of analysis;
- In case of RCT: randomization and allocation, concealment, blinding, intention-to-treat analysis.

### Analysis

Because we expected all kinds of study designs and case reports in different, selected and mostly small populations, among them hardly any RCTs, we estimated that pooling of results would not be feasible. Therefore, the analysis will be descriptive, taking methodological quality into account.

The benefits and risks of state of the art therapeutic interventions in the general population will be summarized and evaluated for the population with ID, taking cognitive, behavioral and motor limitations into account.



## Results

We initially found 1912 studies. In MEDLINE many were labelled clinical trial or review, but most were not actually trials, nor reviews but only descriptions of cases or small case series of syndromes. On the other hand some of the papers not labelled as clinical trial or review appeared to be intervention studies. Appendix 1 lists all papers in which some intervention was described except pharmaceutical trials or comparisons of surgical techniques with people with ID/cerebral palsy.

Initially we did not find any studies on laser assisted techniques for refraction errors. A subsequent search on the use of laser assisted techniques for children yielded five studies.

### Methodological quality

The methodological quality of intervention studies for visual impairment and ophthalmological conditions generally appear poor, both in populations with intellectual disabilities / cerebral palsy and in the general population.

The majority of papers concerning people with intellectual disabilities/ cerebral palsy describe a large diversity of syndromes and conditions and do not involve interventions. Intervention studies appear very scarce for this population and are primarily case reports and small case series. In our selection are one RCT, four case-control studies, nine retrospective follow-up studies (six on keratoconus), and two case series. Three studies with a good case-control design are about eyeglasses and accommodation problems.

Size and power of studies in ID and in the general population are almost always too small. Most studies describe populations of an institution or (for the general population) visiting a clinic and therefore representativeness is not obtained. The studies of Stewart (2005) and Cregg (2001) are the exception, they use a cohort of children with Down syndrome.

Follow-up (sometimes retrospective) is often good, up to ten years or more.

Information on consent procedures, inclusion, participation, reasons for failure is often fragmentary.

The outcome measures are ‘clinical’ and do mostly not focus on relevance for the patient. For example: lower pressure in the eye in cases where the patient does not experience any discomfort. Quality of life outcomes are rarely provided.

Given the poor quality of the research the quality of data analysis cannot be good.

In the few cases of RCTs the information randomization, allocation, blinding and intention to treat analysis is often incomplete.

We rated the quality of the included studies in to systems. In the Sign system (BMJ 2001; 323; 334–336) ranging from 4 = expert opinion to 1++ = high quality meta-analysis or RCTs with very low risk of bias. We rated also according to the grades of recommendations as published in Spine (2007; 32(19S): S66–S72). From the highest 1A = clear risk/benefit to 2C = unclear risk/benefit based on observation. Sign grades of the thirteen included studies: none was in the highest category, one study each in class: 1-, 2+ and 2- and ten studies in the almost lowest class: 3. The Spine grades again not one in the highest grade, one in 1B, the second highest grade; five studies in 1C and seven in 2C, the lowest grade.

General population: the Cochrane reviews concluded in six of thirteen reviews that there was no sufficient evidence to answer the research question.

In the following section we will discuss the included studies. More detailed information on the sixteen selected studies is presented in Appendix 1 and the Cochrane reviews for the general population are listed in Appendix 2.

## Included studies

- Low vision, visual impairment and blindness

### Population with ID or cerebral palsy

One paper on visual training for people with cerebral palsy was found (Duckman, 1980). This describes ten cases of children with cerebral palsy who received training for improvement of visual skills. All children showed significant improvement.

### **General population**

One Cochrane review about reading aids, concluding that more research is needed: especially on the added value of electronic devices for reading compared to optical devices (Virgili, 2006). Another about rehabilitation programs for orientation / mobility which concludes there is not sufficient evidence and RCTs should be conducted to compare different rehabilitation programs (Virgili, 2006).

- **Refractive errors**

#### **Population with ID or cerebral palsy**

A real double masked RCT on effect of spectacles on behavior of persons with ID (Bader et al. 1980) describes positive effects on behavior, motor skills, recognition.

The effect of double focus glasses on visual acuity in children with Down syndrome was investigated by Stewart (2005). The result of this carefully designed case/control study with 34 participants is that the treatment group showed consistent and more accurate accommodation and that compliance with spectacles was high in both groups. Surprisingly, accommodation was not only improved through the bifocal segment but also through the distance part of the lens.

A small second study described successful behavioral interventions to optimize compliance with prescribed glasses in four persons with ID who previously refused to wear glasses. (DeLeon et al., 2008)

There were six papers on correction of refractive errors in children with high myopia or high hypermetropia or anisometropia who could not be treated with spectacles or contact lenses with four years follow-up or more treated with photo refractive keratectomy (PRK) and/or laser-assisted in situ keratomileusis LASIK and/or laser-assisted sub epithelial keratectomy (LASEK). These papers describe case series without controls, or case series comparing the two techniques (n = 74-11). Outcomes are visual acuity and refraction errors at the end of follow-up (Astle, 2008; Yin, 2007; Levenger, 2006; Dvali, 2005 Paysse, 2004; O'Keefe, 2004). In the studies participated patients with neurodevelopmental disorders, this is not clearly described but only mentioned as the cause of treatment failure with glasses or contact lenses. The majority reaches refraction normalization within 2 Dioptres, and almost all children showed improved visual acuity.

Paysse (2003) describes the implementation of an anaesthesia protocol for PRK for non cooperative children and claims this is safe for adults as well.

### **General population**

One Cochrane review compared results of PRK and LASIK procedures in adults, and found only a small difference postoperatively. None of the studies used in this review found adverse effects during follow-up.

For information on feasibility we looked at the compliance for glasses in the general population and found two explicitly on compliance. In a randomized, double masked, crossover trial with 11 year old school children in Japan (n = 92) wearing either single vision lenses or progressive addition lenses a high compliance of 98% (as reported by the children!) was found over an 18 month period (Suemaru, 2008). Castanon Holguin (2006) reports on a cohort of 493 children (5-18 years old) who had received glasses free of charge for refraction errors > 0.5 Dioptres. At a not previously announced check at school within 18 months after providing the glasses, only 13.4% was actually wearing the glasses, an additional 34% had the glasses with them but were not wearing them. Maybe > 0,5 Dioptre is not enough to experience the glasses as useful.

- **Cataract**

### **Population with ID or cerebral palsy**

Goto (1995) describes cataract surgery results in 24 persons with ID and physical handicaps and claims improvement in vision and quality of life in all participants. He concludes that surgery is possible with additional care.

### **General population**

One Cochrane review about mono- or multi focal intraocular lenses reporting that multi focal lenses provide better near vision but have more adverse effects: lower contrast sensitivity and halos (Leyland, 2006). One concluding that anterior vitrectomy and optic capture are effective in bilateral congenital cataract. (Long, 2006). One by Fedorowicz (2006), who compared day-care and in-patient care for age-related cataract surgery and found no clinical difference (but clear financial difference) in outcomes and risks.

- Amblyopia/ strabismus

**Population with ID or cerebral palsy**

One retrospective longitudinal file study of 33 patients with cerebral palsy and strabismus, where strabismus was treated with surgery or training looking back over ten years. Results: strabismus persisted in all three treatment groups (Lo Cascio, 1987).

**General population**

One Cochrane review on treatment of strabismic amblyopia concludes: occlusion, whilst wearing necessary refractive correction, appears to be more effective than refractive correction alone (Shotton, 2008).

Another by Hatt (2006) on the rare condition of amblyopia due to obstruction of the clear passage of light, who found no RCTs.

A third on infantile esotropia (Elliot, 2005) again no evidence found.

The fourth on adjustable versus non adjustable sutures for strabismus surgery (Sundaram, 2005) conclusion: no reliable conclusions could be reached.

The last on interventions for intermittent distance exotropia (Hatt, 2006). could include one randomized controlled trial indicating that unilateral is more effective than bilateral surgery.

- Glaucoma

**Population with ID or cerebral palsy**

One study of Goto (1995) reporting on 24 patients (42 eyes) including ten Down syndrome for cataract surgery with significant improvement in vision and quality of life in all. One eye had serious complication.

**General population**

There are nine Cochrane reviews on glaucoma:

- 1 Acupuncture treatment, no RCT found, no solid evidence found (Law, 2007);
- 2 Laser trabeculoplasty in recently detected open angle glaucoma compared with surgical trabeculoplasty and medication, no clear evidence found (Rolim de Moura, 2007);
- 3 Medical interventions of primary open angle glaucoma. Conclusion: The results of this review support the current practice of IOP (intra ocular pressure) lowering treatment of OHT (ocular hypertension). A visual field protective effect has been clearly demonstrated for medical IOP lowering treatment (Vass, 2007);
- 4 Lens extraction for chronic angle-closure glaucoma. The authors conclude: there is no evidence from good quality randomized trials or non-randomized studies of the effectiveness of lens extraction for chronic primary angle-closure glaucoma (Friedman, 2006);
- 5 Aqueous shunts for glaucoma. Conclusion: To date there is no evidence of superiority of one shunt over another (Minckler, 2006);
- 6 Intra-operative Mitomycin for glaucoma surgery. Conclusion: Mitomycin reduces the risk of surgical failure but there was insufficient power to detect any increase in other serious side effects such as endophthalmitis (Wilkins, 2005).
- 7 Medical versus surgical interventions for open angle glaucoma. Conclusion: for mild OAG, no significant difference between medication or trabeculectomy. For severe OAG no evidence to determine the effectiveness of contemporary medication (prostaglandin analogues, alpha<sub>2</sub>-agonists and topical carbonic anhydrase inhibitors) compared to surgery (Burr, 2005);
- 8 Interventions for normal tension glaucoma. Conclusion: the results for calcium antagonists are promising, but larger trials needed. Irrelevant outcomes for the patient were not taken into account (Syha, 2003);
- 9 Post operative 5-Fluorouracil for glaucoma. Conclusion: 5-Fluorouracil appears to be effective in reducing the likelihood of surgical failure of trabeculectomy (Wormald, 2001).

- Keratoconus

#### **Population with ID or cerebral palsy**

We found six papers comparing the results of corneal grafting for people with Down syndrome and others with keratoconus. They describe retrospectively results of penetrating keratoplastic surgery in the general population, including/

comparing results in people with Down syndrome (Sharif et al., 1991; Völker-Dieben et al., 1993; Haugen et al., 2001; Bodenmueller et al., 2003; Wylegala et al., 2006; Wroblewski, 2006). Results were generally less than for the general populations, one study found serious complications only with penetrating keratoplastic surgery and none with non penetrating techniques. Clear grafts after five years were found in 50-67% of the cases.

### **General population**

No Cochrane reviews on keratoconus were found.

There were many reviews on keratoplastic surgery, none of those mentioned Down syndrome patients in the research population, on the other hand Down syndrome was not an exclusion criterion either. Characteristics of the study population mostly concerned ophthalmological criteria like visual acuity, presence of cataract, and previous eye surgery.

- **Cerebral visual impairment**

### **Population with ID or cerebral palsy**

We found one descriptive file study on rehabilitation in a series of 76 children with cerebral palsy (Lanners, 1999). Patients showed considerable increase in attention and visual curiosity. Six children with severe neuromotor damage showed no change in visual functions.

### **General population**

None was found.

## **Feasibility of interventions**

We integrate the fragmentary information on all studies above with state of the art treatments that are routinely applied in the general population, to assess their proven or supposed feasibility in people with intellectual disabilities.

### **Refractive errors**

Several studies report that compliance with eye glasses is reasonable in people with intellectual disabilities, with up to 80% of prescribed glasses being

regularly worn (Stewart, 2005). For children with Down syndrome fitting glasses can be a problem, because of their facial characteristics. Woodhouse et al., formulated requirements for good fitting spectacles for this group. If glasses are not easily accepted habituation programs can be tried in which glasses are offered in situations that the patient is expected to gain directly (for example by offering the glasses before dinner to make the food more visible for patients who like to eat). DeLeon (2008) describes four persons with severe ID who refused glasses who started wearing the glasses after behavioral interventions. We conclude that this generally accepted treatment is feasible for people with ID.

For refractive errors, laser techniques developed in the past decades seem to open new perspectives. We found one study: "In a retrospective study of eleven children with amblyopia due to high anisometropia, high astigmatism and/ or high myopia and with associated developmental delay. The surgical refractive treatment eliminated or reduced the anisometropia, reduced the astigmatic error, improved vision and improved the daily function of the children with developmental delay. There were no complications or untoward results." (Levenger, 2006).

Photorefractive keratectomy (PRK) and laser-assisted subepithelial keratectomy (LASEK) does not require cooperation of the patient and in children can be performed under general anaesthesia (Astle et al., 2008; Paysse, 2003). Although we did not find any studies on this subject in adults with intellectual disabilities, this may be a promising new therapy for refractive errors in this group.

### **Amblyopia**

There is no evidence that routine procedures for amblyopia treatment (corrective eyeglasses, occlusion) are more difficult to tolerate for children with intellectual disabilities. Therefore, amblyopia treatment in this group should be as usual and studies to monitor the effects are necessary.

### **Cataract**

Cataract is a common finding in people with intellectual disabilities (Splunder et al., 2004). We found one paper specifically on complications of cataract surgery in 24 patients with severe intellectual and physical disabilities (Goto et al., 1995). Two patients had complications in one eye, but not in the other. The authors conclude that surgical treatment in this group is promising if various technical difficulties in the pre- and postoperative stage are anticipated.



There is no evidence that feasibility of cataract surgery in this group is different from that in the general population. Therefore we advise conventional treatment for people with ID with additional pre- and postoperative care if necessary.

### **Amblyopia / Strabismus**

Strabismus is diagnosed in the Netherlands in 42% of the adult population with intellectual disabilities (Splunder et al., 2004). It is unknown why these people had not received treatment as usual. We found no sound evidence against surgical correction of strabismus in patients with intellectual disabilities. Therefore, we conclude that treatment of strabismus in people with intellectual disabilities should be the same as is usual in the general population including surgery, with additional care in the pre- and postoperative period if necessary.

### **Glaucoma**

Glaucoma is clinically 'invisible' and this is even more so for people with ID. Measuring eye pressure is more difficult than in the general population if the patient is not able to cooperate and anxious. Yet it is possible to measure eye pressure and if hypertension is found again there is no evidence against treatment of people with ID the same as usual.

### **Keratoconus**

Keratoconus is a frequent finding in people with Down syndrome. Acute keratoconus in Down syndrome has already been mentioned in 1956 (Hofmann, 1956). Many case reports followed and penetrating keratoplasty has been performed occasionally since. We found six retrospective studies of surgical treatment of keratoconus for people with Down syndrome. The general conclusion is that for people with Down syndrome, surgical results are less positive, but still fairly good with survival rates of a clear graft of 67% or more after five years (see Appendix 1).

Haugen (2001) found specific corneal abnormalities in people with Down syndrome. People with Down syndrome also tend to rub their eyes. If this is noticed examination of the eyes might reveal keratoconus in an early stage.

A factor leading to an increased risk of complications may be that surgery in people with Down syndrome often takes place after exacerbations with extreme hydrops of the cornea. The risk may be diminished if surgery is performed in an earlier stage, before hydrops occurs, because in that case, the diameter of the graft can be smaller which generally leads to fewer complications (Völker-Dieben et al., 1993).

According to Völker-Dieben et al. (1993), surgical results of keratoplastic surgery in Down syndrome can be improved if the following conditions are fulfilled:

- 1 Loss of well-being and functioning as a result of impaired vision;
- 2 Intact posterior segment of the eye;
- 3 No history of self-inflicted trauma or extensive eye rubbing;
- 4 No risk of trauma by others;
- 5 Ability to tolerate eye-patch, protective glasses and eye drops (to be tested before the operation);
- 6 Presence of parent or caretaker who is motivated and able to stay with the patient during the first four postoperative days, to instill eye drops several times daily and to take immediate action when the eye looks red or inflamed.

We conclude that under the above conditions, penetrating keratoplastic surgery is well possible in people with Down syndrome and probably also in other people with intellectual disabilities. Special care should be taken to diagnose keratoconus in an early stage to prevent further damage to the cornea. This way vision loss can probably be prevented and quality of life preserved.

## Discussion

We found that research in the area of visual problems in the population with but also without ID the quality is poor. The majority of papers consists of case series. Another problem maybe that (double) masking is often difficult because interventions are visible. Solutions are sought by researchers for example with 'placebo' (= plano) glasses.

With respect to feasibility we were surprised to find that with ID are never explicitly excluded from studies. Sometimes is even mentioned in a subordinate clause that patients have for example 'neurodevelopmental delay', this is not specified nor mentioned in tables. Only ophthalmic characteristics are mentioned in tables and results, which can be seen as an indication that ID is not considered a problem as long as the ophthalmological problem is treatable. This may explain that we found almost no evidence against 'treatment as usual'. Only for keratoconus less positive results in Down syndrome are mentioned, but still the

majority improves and measures are suggested (and seem feasible) to improve prognosis.

## Conclusions

### Implications for practice

At present 40 % of the visual ophthalmological problems of people with ID in the Netherlands is not diagnosed, and therefore left untreated. We do not expect this to be much different in other high income countries. The literature reviewed in this study shows that existing conventional treatment is possible for people with ID without much increased risks and with benefits on vision and quality of life. It is clear that in case of problem behavior like eye poking or eye rubbing, or high anxiety levels this has to be dealt with and than positive results are feasible. In our recommendations especially for keratoconus surgery we have stressed that extra attention should be given to problems which can be foreseen. We conclude that at present there is no evidence that people with ID should not be diagnosed and treated like anybody else, where necessary with some extra care especially pre and post operative of surgical treatment.

### Implications for research

The body of literature on treatment of visual and ophthalmological problems in people with ID is currently rather small. There are, at present, no indications that the existing treatments and therapies that are common practise for the general population, pose prohibiting problems for persons with ID. The special attention required for this population is well known and could be easily included in treatment for this special group.

Be this as it may, hard evidence on effectiveness, feasibility, safety and long term effects and side effects is, in our view, urgently required. Without such evidence, the common practise of being rather safe than sorry will deprive a considerable number of people in our societies of care and treatments that could definitely improve their quality of life, and at the same time make the work of staff and carers in the field of ID more fruitful and satisfying instead of unduly burdened.

Against this background we would like to make a case for a series of long-term, prospective well controlled RCT studies on the treatment of a.o. refractive

errors, strabismus, amblyopia, cataract, glaucoma and keratoconus with surgical, non-surgical, behavioral and training methodologies. The driving philosophy, and the road to funding, is that people with ID deserve the same level of health care as any one else.

Any one working with people with ID knows that the people that are entrusted to their care are absolutely worth their while. The same way that color vision deficiencies gave unique insights into human vision, the care for, and the research pertaining to people with intellectual disabilities gives a unique view on human nature, brain function and consciousness.

## Appendix 1

### Characteristics of included studies in the population of people with ID and cerebral palsy

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<b>Refractive error</b>	<b>The effects of corrective lenses on various behaviors of mentally retarded persons</b>
Authors	Bader, D. and W.E. Woodruff, 1980
Methods	Behavioral study on the effect of glasses for persons with ID. Prospective controlled clinical trial. One intervention group and three control groups. Behavior was assessed with an adapted version of the Adaptive Behavior questionnaire (Heber, 1961). Main hypothesis was tested with $\chi^2$ statistic; further analysis of scores with ANOVA. Follow-up 2, 4 and 8 weeks.
Participants	287 persons with ID in three age groups: 0-6 years (115), 7-17 years (71) and > 18 years (101). ID levels across age groups were: Borderline (6.6%), Mild (25.8%), Moderate (40.8%), Severe (19.2%) and Profound (7.3%).
Interventions	The 3 age groups were sub-divided in 4 intervention groups (# patients per age group): (a) <i>treatment</i> , received new spectacles (21/21/33), (b) <i>delay</i> , received new glasses after 2 months (17/16/23), (c) <i>no spectacles required</i> (60/17/26) and (d) <i>placebo</i> (17/17/19), persons that did not need correction but were given plano glasses.
Outcomes	Questionnaire scores: social behavior, gross- and fine motor behavior, picture/object recognition, writing, and objective and subjective assessment of feelings about the effects of spectacles. The treatment group showed significant improvements on all categories; the youngest age group (0-6 yrs) showed more improvement than the other age groups. Follow-up: % of patients wearing glasses in <i>treatment/placebo</i> groups after 2 weeks: 81.4/81.2, 4 weeks: 82.7/77.4 and 8 weeks: 81.4/66.1
Notes	none
Sign classification	1-
Spine grade	1B

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<b>Refractive error</b>	<b>Accommodation and refractive error in children with Down syndrome: cross-sectional and longitudinal studies</b>
Authors	Cregg, M.J., et al., 2001
Methods	Small prospective intervention study along side a large cross-sectional and longitudinal population based study.
Participants	75 people with Down syndrome
Interventions	Glasses

Outcomes	Accommodation error was stable over time, even correction with glasses left under-accommodation unchanged
Notes	Good follow-up rate, loss 1 because of death, 3 moved elsewhere
Sign classification	3
Spine grade	2C

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**Refractive error    In focus: the use of bifocal spectacles with children with Down syndrome**

Authors	Stewart, R.E. et al., 2005
Methods	Comparative non-randomized interventional study with follow-up. Accommodation response was measured with dynamic retinoscopy. Compliance with, and impact of, bifocal spectacles was evaluated with a simple questionnaire. Three follow-ups over a period of 5 months. Statistical analysis involved Student's t and $\chi^2$ tests, two-way ANOVA and various subsequent post hoc tests.
Participants	34 children with Down syndrome (age 5-11 years) divided in two equal, matched <i>treatment</i> and <i>control</i> groups.
Interventions	The treatment group was prescribed bifocal spectacles with a +2.5 D addition; the control group was given single vision lenses to correct any clinically significant refractive error.
Outcomes	Group matching: no significant difference between groups was found at study onset (Student's t and $\chi^2$ tests; $p < 0.05$ ). The treatment group showed consistent and more accurate accommodation ( $p < 0.5$ ). Compliance with spectacles was high in both groups ( $> 82\%$ ).
Notes	Accommodation was improved both through the bifocal segment and, surprisingly, also through the distance part of the lens.
Sign classification	2+
Spine grade	1C

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**Eyeglasses    Increasing wearing of prescription glasses in individuals with mental retardation**

Authors	DeLeon, I.G., et al., 2008
Methods	Evaluation of an intervention for promoting wearing of prescription glasses, through partial component analysis.
Participants	4 persons with ID who did not wear their prescribed glasses
Interventions	Distraction through non-contingent reinforcement (NCR), in combination with response cost and brief response blocking.
Outcomes	All four learned to wear glasses.
Notes	none
Sign classification	3
Spine grade	2C

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<b>Cataract</b>	<b>Intraocular lens implantation in severely mentally and physically handicapped patients</b>
Author	Goto, S. et al., 1995
Methods	Comparison with implants in conventional patients.
Participants	The study population consisted of 13 severely mentally and physically handicapped patients (21 eyes), 10 patients with Down syndrome (19 eyes), and a patient suffering from encephalo-myopathy (2 eyes). A total of 42 eyes of 24 patients.
Interventions	Posterior chamber intraocular implantation (PC-IOL) under general anaesthesia.
Outcomes	Significant improvement in vision and quality of life. Severe postoperative complications occurred in one eye.
Notes	PC-IOL in patients with severe mental and physical handicaps should be considered as an appropriate and promising application in spite of the various technical difficulties in the pre- and postoperative management of these patients.
Sign classification	3
Spine grade	1C

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<b>Strabismus</b>	<b>Treatment for strabismus in cerebral palsy</b>
Authors	Lo Cascio, G.P., 1987
Methods	Retrospective longitudinal file study of patients with cerebral palsy and strabismus, where strabismus was treated with surgery or training.
Participants	33 patients with cerebral palsy. Most of them were spastic. Divided into 3 groups (# patients): <i>surgery</i> (11), <i>training</i> (9), and <i>no intervention</i> (13). The averages were 7.2, 10.0 and 10.7 years, respectively.
Interventions	Surgery was performed at ages ranging from 1 to 18 years, in the period 1967 to 1981. Procedures included muscle recession, resection, supraplacement, tenectomy and myomectomy. Training included sensory and motor tasks and used requirements such as filters, stereoscopes. Training duration was between 4 and 9 months for 7 of the 9 patients in this group; training lasted 1 month in one case and 22 months for one patient.
Outcomes	Tropia eso and exo magnitudes were determined before and after treatment. Follow-up data for <i>surgery</i> were grouped in periods 1-5 and 6-14 years after treatment. The patients in the <i>no intervention</i> group were examined between 1970 and 1986, with an interval of at least 10 years between initial and final examination. Results: average pre- and post treatment magnitudes of tropia $\Delta$ (prism diopters) in the three groups are: <i>surgery</i> : 43/45, <i>training</i> 41/37, and <i>no intervention</i> 42/51. As measurement accuracy is 8 $\Delta$ there is no significant

	difference between treatments. Further, strabismus persisted in all 3 treatment groups.
Notes	The author evaluates his 19 years experience with his own training methodology.
Sign classification	2-
Spine grade	1C

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**Keratoconus Long-term graft survival in patients with Down syndrome after penetrating keratoplasty.**

Authors	Wroblewski, K.D., et al., 2006
Methods	Retrospective file study with follow-up of 4-88 months combined with visual function assessment at the end
Participants	Patients with Down Syndrome who underwent keratoplastic surgery for keratoconus. Between 1987-98: 13 patients (18 eyes) were found in the files
Interventions	Keratoplastic surgery for keratoconus in people with Down syndrome.
Outcomes	Clarity of graft: in 10 eyes, 3 cloudy, 3x second surgery had been necessary
Notes	none
Sign classification	3
Spine grade	2C

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**Keratoconus Corneal grafting for keratoconus in mentally retarded patients**

Authors	Haugen, O.H. et al., 2001
Methods	Retrospective over 1974-2000 study of files of patients with keratoconus who underwent one of 3 types non-penetrating, lamellar keratoplasty or epikeratophakia or penetrating keratoplasty.
Participants	41 patients with ID mean age 36.7 ( $\pm$ 10.8)
Interventions	One of 3 types non-penetrating, lamellar keratoplasty or epikeratophakia or penetrating keratoplasty.
Outcomes	All serious complications were after penetrating keratoplasty (PK), graft failure in 44% PK en in 16% NP; more problems at age > 40. 5 years; graft survival 74,9%; visual acuity improvement in all cases post operative, but later deterioration.
Notes	none
Sign classification	3
Spine grade	2C

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**Keratoconus Penetrating kerastoplasty for keratoconus: complications and long-term success**

Authors	Sharif, K.W. and T.A. Casey, 1991
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Methods	Retrospective file study with follow-up minimum 4 years, average 6.1 years
Participants	100 participants, age 11-51 years, of which 4 with Down syndrome. Participants had no previous intraocular surgery; follow-up at least 4 years. Surgery was performed on people with Down syndrome only after they had repeated attacks of acute hydrops.
Interventions	Keratoplasty; technique evolved during period of study.
Outcomes	Long term prognosis is favorable: a clear cornea resulted 93% of the participants. 4% remained blind. Two of the patients with Down syndrome developed bacterial keratitis, one a fungal keratitis. Long term treatment with steroids.
Notes	All participants with Down Syndrome showed eye rubbing before treatment.
Sign classification	3
Spine grade	1C

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<b>Keratoconus</b>	<b>Amniotic membrane transplantation with cauterization for keratoconus complicated by persistent hydrops in mentally retarded patients</b>
Authors	Wylegala, E. and D. Tarnawska, 2006
Methods	Retrospective file study. Pearsons r coefficient was used to correlate initial corneal protuberance and final corneal thickness and time of resolution; $p < 0.05$ . Follow-up 6-28 months.
Participants	10 patients with ID (13-37 years), 8 of which had Down syndrome, that suffered from refractory hydrops for more than 6 months.
Interventions	Amniotic membrane transplantation with cauterization for keratoconus for persistent hydrops of the cornea in mentally retarded patients. Cauterisation and stromal puncture, adding of amniotic membrane, followed by 3 weeks local fluoroquinolone 4xdd and Beta blocker 2xdd. Follow up of 6-28 months. Complications 3x surgery because of loose suture, 1x delayed epithelialisation, 1x corneal vascularisation
Outcomes	Difference in corneal protuberance, final corneal thickness and time of resolution Corneal protuberance disappeared after $21.4 \text{ days} \pm 9.5 \text{ days}$ in all eyes. Corneal thickness was back to $0.47 \pm 0.11 \text{ mm}$ after three weeks. Symptoms of eye discomfort disappeared 3 weeks after surgery for 3 patients, was diminished in 3 and unchanged in 4. For the latter 7 patients eye discomfort had disappeared after 2 months. 2 diminished corneal thickness en 1 hard to say because of functioning
Notes	Complications during surgery were: suture undone 3 times, delayed epithelialisation once and corneal vascularisation once.
Sign classification	3
Spine grade	2C

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<b>Keratoconus</b>	<b>Surgical treatment of corneal pathology in patients with Down syndrome</b>
Authors	Völker-Dieben. H.J. et al., 1993
Methods	Patients with keratoconus were selected from a retrospective survey of corneal transplants performed between 1976 and 1990. This yielded 268 cases, out of a total of 1748. Graft survival time was calculated using the actuarial life table method of Petro et al. (1976)
Participants	There were 268 patients with keratoconus, 29 of which had Down syndrome. Mean age of the latter group was 35.5 years, range 8 – 61. Grafts on patients with Down syndrome (n = 33) and general keratoconus population (n = 238)
Interventions	Corneal transplant with the method Harms & Mackensen, followed by corticosteroid en antibiotic eyedrops for at least one year.
Outcomes	The criterion for graft survival was clarity. Graft survival between 1 and 5 years ranged from 85 to 67% in the patients with Down syndrome <i>vs</i> from to 99-90% in the 238 non-Down. The main causes for graft failure in the Down syndrome group <i>vs</i> non-Down patients were bacterial infection (9.7% / 1.3%) and trauma (6.5% / 0%).
Notes	none
Sign classification	3
Spine grade	2C
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<b>Keratoconus</b>	<b>Perforierende Keratoplastik bei Trisomie 21</b>
Authors	Bodenmueller, M. et al., 2003
Methods	Retrospective file study from 1985 – 2002. Follow-up 2 – 15 years; average 7 years.
Participants	9 patients (16 eyes) with Down syndrome with keratoconus An age-matched control group of 19 non-Down syndrome patients (25 eyes) with keratoconus. Average age 35 years.
Interventions	Penetrating keratoplasty (PK)
Outcomes	For the <i>Down syndrome patients</i> , 5 grafts (out of 16) were clear at the last follow-up. In 7 eyes at least 1 re-PK had to be performed. Seven eyes developed graft rejection and 4 eyes needed conjunctival grafting because of non-healing epithelial defects. Ten eyes developed cataracts and 3 secondary glaucoma. In the <i>control group</i> 22 out of 25 operated corneas were clear at last follow-up.
Notes	The poor results for the Down syndrome patients are not only caused by lack of cooperation of patients but also by the high rate of emergency procedures (3/16), presence of lid anomalies and very advanced keratoconus.
Sign classification	3
Spine grade	2C
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### Cerebral visual impairment

#### Early intervention for children with cerebral visual impairment: preliminary results

Authors	Lanners, J. et al., 1999
Methods	Re-assessment of the clinical history of patients over 3 years and review of the relevant literature. Visual impairment was assessed through observation of behavior in daily life circumstances.
Participants	76 patients with CVI; ages 7 months – 4 years, mean 2.4 years. 30 patients were rehabilitated sufficiently to allow assessment of visual function improvement.
Interventions	Psychomotor, sensory, cognitive and psychological, including visual rehabilitation.
Outcomes	Visual Development Scale. 20 out of 30 patients showed considerable increase in attention and visual curiosity. 6 children with severe neuromotor damage showed no change in visual functions. The results confirm the general findings in the literature.
Notes	This study is a 'self-re-assessment' of treatment practice in the authors' institution.
Sign classification	3
Spine grade	2C

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### Visual impairment

#### Effectiveness of visual training on a population of cerebral palsied children

Author	Duckman, R.H. 1980
Methods	An experimental program of visual training was carried out. No statistical analysis but minute descriptions of each case. Evaluation of effects after 1 year, by comparing pre- and post training scores.
Participants	10 children with cerebral palsy selected from of 25 inpatients with visual problems. Inclusion criteria: intellectual capacity to follow instruction, sufficient motor control to participate in prescribed activities, at least one more year stay in hospital. Ages: from 1 – 11 years; IQ between 70 and 112.
Interventions	Training 30 min per day, 5 days a week during 10 months. The objective was improvement of visual- and other skills that pertain to learning. Training included: fixation, ocular motilities, eye pursuit movements, hand-eye coordination, accommodative loose lens rock, forced convergence training, form- and size matching.
Outcomes	All children showed significant improvements in ocular motility, 3 triopas disappeared spontaneously, improved accommodative facility and flexibility in 2/3 of the children.
Notes	Evaluations were not independent (performed by staff). Nevertheless, the results seem convincing.
Sign classification	3
Spine grade	1C

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## Appendix 2

### Details of Cochrane reviews

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<b>Shotton</b>	<b>2008</b>
Strabismic Amblyopia	Interventions for strabismic amblyopia
Nr of included studies	2 RCTs
Total nr of participants	64
Sufficient evidence to answer study question	Yes, partly
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Author's conclusions	<p>Occlusion, whilst wearing necessary refractive correction, appears to be more effective than refractive correction alone in the treatment of strabismic amblyopia. Combining occlusion and refractive correction with near activities may be more effective than occlusion and refractive correction alone. Further study of the role of near activities is necessary before a more definitive conclusion can be made.</p> <p>Results of a full trial are expected within the coming year. No RCTs were found that assessed the role of either partial occlusion or optical penalisation to refractive correction for strabismic amblyopia.</p>
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<b>Law</b>	<b>2007</b>
Glaucoma	Acupuncture for glaucoma
Nr of included studies	None
Total nr of participants	0
Sufficient evidence to answer study question	No
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Author's conclusion	<p>At this time, it is impossible to draw reliable conclusions from the available data to support the use of acupuncture for the treatment of glaucoma. Since most glaucoma patients currently cared for by ophthalmologists do not use non-traditional therapy, the clinical practice decisions will have to be based on physician judgement and patients' value given this lack of data in the literature.</p>
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<b>Rolim de Moura</b>	<b>2007</b>
Glaucoma	Laser trabeculoplasty for open angle glaucoma
Nr of included studies	19
Total nr of participants	2137
Sufficient evidence to answer study question	Not clearly
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**Author's conclusion** Evidence suggests that, in people with newly diagnosed OAG, the risk of uncontrolled IOP is higher in people treated with medication used before the 1990s when compared to laser trabeculoplasty at two years follow up. Trabeculoplasty is less effective than trabeculectomy in controlling IOP at six months and two years follow up. Different laser technology and protocol modalities were compared to the traditional laser trabeculoplasty and more evidence is necessary to determine if they are equivalent or not. There is no evidence to determine the effectiveness of laser trabeculoplasty compared to contemporary medication (prostaglandin analogues, topical anhydrase inhibitors and alpha2-agonists) and also with contemporary surgical techniques. Also there should be further investigation into the effectiveness of laser trabeculoplasty in specific racial groups, specific diagnostic groups, such as pseudoexfoliation and pigmentary glaucoma and different stages of OAG. More research is also required determining cost-effectiveness of laser trabeculoplasty in the management of glaucoma.

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**Virgili** **2006**  
 Low vision Reading aids for adults with low vision  
 Nr of included studies 8 cross-over + 1 three parallel-arm  
 Total nr of participants 221 + 243 = 464  
 Sufficient evidence to answer study question No

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**Author's conclusion** Further research is needed on the comparison of different types of low vision aids. It will be also necessary to delineate patient's characteristics that predict performance with costly electronic devices as well as their sustained use in the long term compared to simpler and cheaper optical devices.

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**Leyland** **2006**  
 Cataract Multifocal versus monofocal intraocular lenses after cataract extraction  
 Nr of included studies 10  
 Total nr of participants ?  
 Sufficient evidence to answer study question Yes

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**Author's conclusion** Multifocal IOLs are effective at improving near vision relative to monofocal IOLs. Whether that improvement outweighs the adverse effects of multifocal IOLs will vary between patients. Motivation to achieve spectacle independence is likely to be the deciding factor.

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<b>Riaz</b>	<b>2006</b>
Cataract	Surgical interventions for age-related cataract
Nr of included studies	17
Total nr of participants	9627
Sufficient evidence to answer study question	Yes
<b>Author's conclusion</b>	This review provides evidence from seven RCTs that phacoemulsification gives a better outcome than ECCE with sutures. We also found evidence that ECCE with a posterior chamber lens implant provides better visual outcome than ICCE with aphakic glasses. The long term effect of posterior capsular opacification (PCO) needs to be assessed in larger populations. The data also suggests that ICCE with an anterior chamber lens implant is an effective alternative to ICCE with aphakic glasses, with similar safety. Phacoemulsification provides the best visual outcomes but will only be accessible to the poorer countries if the cost of phacoemulsification and foldable IOLs decrease. Manual small incision cataract surgery provides early visual rehabilitation and comparable visual outcome to PHACO. It has better visual outcomes than ECCE and can be used in any clinic that is currently carrying out ECCE with IOL. Further research from developing regions are needed to compare the cost and longer term outcomes of these procedures e.g. PCO and corneal endothelial cell damage.
<b>Friedman</b>	<b>2006</b>
Glaucoma	Lens extraction for chronic angle-closure glaucoma
Nr of included studies	2 non randomized comparative studies
Total nr of participants	48 patients 59 eyes
Sufficient evidence to answer study question	No
<b>Author's conclusion</b>	There is no evidence from good quality randomized trials or non-randomized studies of the effectiveness of lens extraction for chronic primary angle-closure glaucoma.
<b>Virgili</b>	<b>2006</b>
Low vision	Orientation and mobility training for adults with low vision
Nr of included studies	2
Total nr of participants	88 elderly people > 58 years
Sufficient evidence to answer study question	No

**Author's conclusion**      The review found two small trials with similar methods, comparing (O&M) training to physical exercise, which were unable to demonstrate a difference. Therefore, there is little evidence on which type of orientation and mobility training is better for people with low vision who have specific characteristics and needs. Orientation and mobility instructors and scientists should plan randomized controlled studies to compare the effectiveness of different types of (O&M) training. A consensus is needed on the adoption of standard measurement instruments of mobility performance which are proved to be reliable and sensitive to the diverse mobility needs of people with low vision. For this purpose, questionnaires and performance-based tests may represent different tools that explore people with low vision's subjective experience or their objective functioning, respectively.

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**Hatt**                                **2006**  
 Exotropia                            Interventions for intermittent exotropia  
 Nr of included studies            1  
 Total nr of participants        36 + 68 in non randomized control group  
 Sufficient evidence to  
 answer study question        Yes

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**Author's conclusion**      The available literature consists mainly of retrospective case reviews which are difficult to reliably interpret and analyze. The one randomized trial included found unilateral surgery more effective than bilateral for basic intermittent exotropia but there remains a need for more carefully planned clinical trials to be undertaken to improve the evidence base for the management of this condition.

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**Long**                                **2006**  
 Cataract                              Surgical interventions for bilateral congenital cataract  
 Nr of included studies            4  
 Total nr of participants        65+16+6+50 = 137  
 Sufficient evidence to  
 answer study question        Yes

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**Author's conclusion**      Evidence exists for the care of children with congenital or developmental bilateral cataracts to reduce the occurrence of visual axis opacification. Further randomized trials are required to inform modern practice about other concerns including the timing of surgery, age for implantation of an intraocular lens and development of long-term complications such as glaucoma and retinal detachment.

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<b>Minckler</b>	<b>2006</b>
Glaucoma	Aqueous shunts for glaucoma
Nr of included studies	15
Total nr of participants	1153
Sufficient evidence to answer study question	No
<b>Author's conclusion</b>	Relatively few randomized trials have been published on aqueous shunts and methodology and data quality among them is poor. To date there is no evidence of superiority of one shunt over another.
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<b>Wilkins</b>	<b>2005</b>
Glaucoma	Intra-operative Mitomycin C for glaucoma surgery
Nr of included studies	11
Total nr of participants	698
Sufficient evidence to answer study question	Yes, not for side effects
<b>Author's conclusion</b>	Intraoperative MMC reduces the risk of surgical failure in eyes that have undergone no previous surgery and in eyes at high risk of failure. Compared to placebo it reduces mean IOP at 12 months in all groups of participants in this review. Apart from an increase in cataract formation following MMC, there was insufficient power to detect any increase in other serious side effects such as endophthalmitis.
<hr/>	
<b>Burr</b>	<b>2005</b>
Glaucoma	Medical versus surgical interventions for open angle glaucoma
Nr of included studies	4
Total nr of participants	888
Sufficient evidence to answer study question	Yes, not for contemporary medication
<b>Author's conclusion</b>	Evidence from one trial suggests, for mild OAG, that the risk of glaucoma progression up to five-years is not significantly different whether treatment is initiated with medication or trabeculectomy. Reduced vision, cataract and eye discomfort are more likely with trabeculectomy. There is some evidence, for more severe OAG, that initial medication (pilocarpine, now rarely used as first line medication) is associated with a greater risk of glaucoma progression than surgery. Surgery lowers IOP more than medication. There was no evidence to determine the effectiveness of contemporary medication (prostaglandin analogues, alpha2-agonists and topical carbonic anhydrase inhibitors) compared with surgery in severe OAG, and in people of black ethnicity. More research is required.

<b>Sycha</b>	<b>2003</b>
Glaucoma	Interventions for normal tension glaucoma
Nr of included studies	8 Only three studies focussed on patient relevant outcomes
Total nr of participants	145+28+52+36+20+?+13+18 = >310
Sufficient evidence to answer study question	No
Author's conclusion	In one study the effect of intraocular pressure lowering on visual field outcome was only significant when data were corrected for cataract development. The results for calcium antagonists are promising, but larger trials have to be performed. Studies that focussed on reduction of intraocular pressure or haemodynamic variables are not necessarily relevant for the outcome in people with normal tension glaucoma.
<b>Wormald</b>	<b>2001</b>
Glaucoma	Postoperative 5-Fluorouracil for glaucoma surgery
Nr of included studies	9
Total nr of participants	614
Sufficient evidence to answer study question	Yes
Author's conclusion	Postoperative injections of 5-Fluorouracil are now rarely used as a planned series but are increasingly used on an ad hoc basis. This presumably reflects an aspect of the treatment that is unacceptable to both patients and doctors. None of the trials reported on the participants' perspective of care which constitutes a serious omission for an invasive treatment such as this.
<b>Fedorowicz</b>	<b>2005</b>
Cataract	Day care versus in-patient surgery for age-related cataract
Nr of included studies	2
Total nr of participants	1284
Sufficient evidence to answer study question	Yes
Author's conclusion	This review provides some evidence that there is a cost saving but no significant difference in outcome or risk of postoperative complications between day care and in-patient cataract surgery. This is based on one detailed and methodologically sound trial conducted in the developed world. The success, safety and cost-effectiveness of cataract surgery as a day care procedure appear to be acceptable but additional well-designed trials are required to confirm these perceptions.

<b>Elliott</b>	<b>2005</b>
Esotropia	Interventions for infantile esotropia
Nr of included studies	0
Total nr of participants	0
Sufficient evidence to answer study question	No
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Author's conclusion	The main body of literature on interventions for IE are either retrospective studies or prospective cohort studies. It has not been possible through this review to resolve the controversies regarding type of surgery, non-surgical intervention and age of intervention. There is clearly a need for good quality trials to be conducted in these areas to improve the evidence base for the management of IE.
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<b>Sundaram</b>	<b>2005</b>
Strabismus	Adjustable versus non-adjustable sutures for strabismus
Nr of included studies	0
Total nr of participants	0
Sufficient evidence to answer study question	No
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Author's conclusion	No reliable conclusions could be reached regarding which technique (adjustable or non-adjustable sutures) produces a more accurate long-term ocular alignment following strabismus surgery or in which specific situations one technique is of greater benefit than the other. High quality randomized controlled trials are needed to obtain clinically valid results and to clarify these issues.
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# 4

## **Is concept-mapping an effective method of introducing complex interventions into intellectual disability services?**

Sjoukes L, Harteloh PPM & Evenhuis HM

## Abstract

As many adults with intellectual disabilities (ID) have an increased risk of low vision, intellectual disability service providers should play an active role in vision rehabilitation. However, low vision rehabilitation is a new and complex intervention for many providers in this field. To increase awareness of vision rehabilitation it was proposed that the process of concept-mapping be tested to see if it would lead to increased acceptance of vision rehabilitation. In our test, the use of concept-mapping appeared to be a satisfactory method for increasing awareness of professionals and middle managers of the complexity of vision rehabilitation, but it was found that it could only be useful for change, if connected to a strategic decision by top management and the adoption of an action plan by the service provider that promoted vision rehabilitation.

## Background

Dutch adults with intellectual disabilities (ID) appear to have a ten times increased risk of visual impairment and blindness, as compared to the general Dutch population aged 55 years and over (Splunder et al., 2005; Klaver et al., 1998). Even young adults with mild ID have a risk comparable to the risk of older adults in the general population. Because many adults with ID make use of residential, day activity, and ambulatory ID services, such services are important factors in low vision rehabilitation. Apart from prescribing glasses and ophthalmological interventions, rehabilitation includes modules that are often innovative yet costly to ID services, such as low vision training of carers, and technical adaptations aimed at better visibility of the living environment and nearby materials. Several disciplines can be involved, with carers as a central group. Therefore, low vision rehabilitation is a new and complex intervention for the ID field.

### Specific aims

To address low vision, we explored whether the method of concept-mapping (Trochim, 1989) is an effective way to introduce low vision rehabilitation into ID services.

## Method

Three ID service providers delivering residential, day care, and sheltered workshop services consented to participate in the intervention study. In addition, three low vision district centers were prepared to provide vision screenings based on IASSID consensus guidelines (Evenhuis & Nagtzaam, 1998). Individual rehabilitation advisories were prepared in cases of visual impairment, including medical (glasses, ophthalmological treatment), environmental (improvement of light, color and contrast, routing), professional (training of staff), and educational advisories (information of clients and families). To combine the views of all concerned disciplines on the execution of such advisories and expected bottlenecks, multidisciplinary groups were formed in each ID service, consisting of carers in homes and day activity centers, middle and top management, physicians, behavioral, educational, and technical staff, and staff of the low vision expertise centers.

We chose the method of concept-mapping (Trochim, 1989) to gather and sort ideas in a conceptual framework. Concept-mapping is a qualitative method, supported by quantitative software and based on principal component analysis. It involves the sorting of similarities, multidimensional scaling, and cluster analysis. We did this in two sessions. The first was a brainstorming session to gather factors expected to influence execution of low vision advisories. Individual rating and clustering by the participants and analysis resulted in clusters of all factors for each ID service. During the second session, each group discussed and interpreted contents of the clusters and named them. Finally, recommendations on conditions that should be realized before introduction of the interventions was formulated and brought to the attention of the top management. After the completion of diagnostic and advisory procedures and training of carer teams, the teams' awareness, appreciation and execution of rehabilitation advisories were measured by means of questionnaires, completed at intervals of 4, 8 and 12 months after receipt of the rehabilitation advisories.

## Findings

**Concept-mapping:** The efforts of 49 participants in the three ID services (40 professional staff, seven middle managers, and two top managers) resulted in the clusters shown in Table 4.1.

The participants generally felt that the clusters reflected relevant themes in their organizations and became aware of the complexity of the innovation. Most factors concerned basic conditions for change: organization (multidisciplinary expertise, competing priorities, planning, support within the organization, embedding in routines, technical and financial organization, communication and evaluation, responsibility and coordination, transfer and evaluation of the rehabilitation advice), budget (financial framework), information (introduction and communication of advisory, involvement of personnel), and quality (quality of advisory, attention to individual client, client autonomy).

*Actions:* Consultation with the top management in the three ID services led to changes in communication procedures, allocation of budget for technical adaptations, and more structured collaboration between disciplines. The three low vision centers synchronized the contents of their advisories and of the training of carers.



Table 4.1  
Factors Influencing execution of low vision rehabilitation  
in intellectual disability (ID) services

ID service 1 Cluster	Rating	ID service 2 Cluster	Rating	ID service 3 Cluster	Rating
Expertise	4.4	Expertise of caregivers	3.8	Multidisciplinary expertise	4.0
		Efficacy expertise diagnosis/ treatment	3.8		
Planning	3.9				
Competing priorities	3.3				
Support in ID service	3.8	Embedding in ID service	4.1	Embedding in ID service	4.0
Financial framework	4.0	Technical/ financial organization	3.6		
Transfer/ evaluation of advice	4.5	Introduction/ communication of advice	4.1	Communication/ evaluation	4.2
		Continuity and teamwork	3.8	Involvement of personnel	4.1
		Understanding/ autonomy of client	4.2	Responsibility/ coordination	4.1
				Quality of advice	3.8
				Individual client	3.7

*Execution of interventions:* Sixty-one adults with ID were identified with heretofore undiagnosed visual impairment or blindness and got expert advice for treatment or rehabilitation (17 received eyeglass prescriptions, 5 received referrals for cataract surgery, and 48 received personalized advisories on low vision rehabilitation, including specific support by carers (i.e., communication, activities, how to use other senses, places and routing, use of light) and technical adaptations). It appeared that during a year of follow-up, four out of seventeen eyeglass prescriptions were still effective and two out of five cataracts were surgically treated. At the end of the year, carers were aware of the personalized rehabilitation advisories in 34/48 cases (71%), advisories were considered useful in 26/61 (43%) and were included in the participant's care plan in 29/61 cases (47.5%). Specific low vision support by carers, as instructed in the training sessions, was actually given in 26/48 cases (54%). Of the technical advisories concerning illumination and contrasts, half were reported completely or partially executed at the end of follow-up.

## Discussion

This study shows that concept-mapping in itself is not effective to support successful introduction of changes into an organization. It appeared satisfactory as a way of increasing awareness of professionals and managers for conveying the fact that low vision rehabilitation is a new and complex intervention for intellectual disability services, requiring change of routine in several involved disciplines and at different levels in the organization. We found that it made professional staff aware that organizational aspects are involved which are outside their sphere of influence or responsibility. However, the identification of factors and clusters has been intuitive, and the clusters do not indicate elementary decision points. As a consequence, resulting actions were primarily operational and ad hoc, and did not change strategic policies of the involved organizations. As a result, the large group of carers and their middle managers who are central actors to the innovation have been insufficiently motivated to change by the top management, and implementation of low vision rehabilitation has been poor.

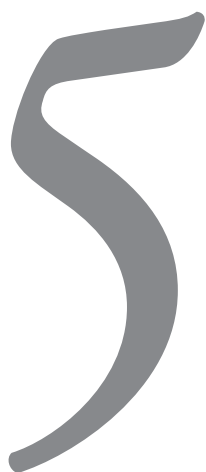
Our conclusion is that concept-mapping may be a good method of increasing awareness among professionals and middle managers of the complexity of an innovation, but it can only be useful to introduce new and complex interventions into ID services if connected to a strategic action plan and decision making by the organization.

## Acknowledgements

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**Rehabilitation of low vision  
in adults with intellectual  
disabilities:**

**The influence of staff**

Sjoukes L, Kooijman AC, Koot JM & Evenhuis HM

## Abstract

**Background** We explored to what extent carers act on treatment and rehabilitation advice for low vision in intellectual disability (ID) services and how this relates to their familiarity with and appreciation of the advice.

**Methods** We followed prospectively sixty adults with a recent diagnosis of low vision during 12 months after their individual intervention advice. Questionnaire scores of carers' familiarity and appreciation of the advice and the quality of implementation were analyzed using Spearman's correlation.

**Results** Familiarity with the advice was reasonable or good in 41/60, appreciation was positive in 34/60, and implementation was reasonable to complete in 20/60 cases. Familiarity, appreciation and implementation were not significantly related to severity of ID or severity of visual impairment. Implementation was positively correlated with familiarity (Spearman's rho 0.602,  $p < 0.001$ ) but not with appreciation (Spearman's rho 0.224,  $p = 0.086$ ).

**Conclusion** It is important to keep professional carers knowledgeable and informed to improve the quality of low vision rehabilitation.

## Introduction

Epidemiological research in the Netherlands has shown that the risk of visual impairment and blindness is ten times higher for adults with intellectual disabilities (ID) aged eighteen years or over than in the general population aged 55 years and over (Splunder et al., 2006). Treatable ophthalmological conditions can be diagnosed in many cases, of which refractive errors and cataracts are the most prevalent (Splunder et al., 2004). Dutch low vision centers are experienced in on-site diagnostic assessment of people with ID, with ophthalmological referral, technical advice on illumination, color and contrast of living quarters and training of carers. However, in clinical practice we noted that advised interventions were not always carried out. Because carers appear central actors in the implementation, we explored:

- To what extent carers are familiar with the given advice;
- How they appreciate the advice;
- To what extent the advice is carried out (implemented);
- How implementation is related to the carers' appreciation of the advice.

## Materials and methods

### Study design and procedure

In a prospective descriptive design we followed adults with ID and a recent diagnosis of low vision during twelve months after advice for treatment and/or rehabilitation. The carers' familiarity with, appreciation and realization of advice were assessed with questionnaires. We emphasized that the same carer should complete the questionnaires at four ( $t_1$ ), eight ( $t_2$ ) and twelve months ( $t_3$ ).

### Study population

Participants were clients of three ID Service providers in the Netherlands, offering centralized and community-based residential and day care. In these services a screening by district low vision centers was planned during 2002–2004. All clients identified with a vision problem and aged eighteen years or over were asked for informed consent to participate in the study. The optometrist of the low vision center classified the severity of the vision problem according to criteria of the World Health Organization (1980): low vision (with glasses if the participant

was already wearing them) if the visual acuity was  $\geq 0,05$  and  $< 0.3$  or blindness if the visual acuity was  $< 0.05$ .

Clients with subnormal vision with a visual acuity  $\geq 0.3$  and  $< 0.5$ , who showed improvement of visual acuity of 0.3 or more with glasses were also asked to participate.

The severity of ID was retrieved from existing client files as assessed with different instruments.

### **Treatment and rehabilitation**

Experts of the low vision centers wrote individual advice for treatment and rehabilitation after behavioral assessments and technical checks of the living environment. Participants and their family were informed about the advice. The low vision center trained the carer team around each participant on-site. Training involved information on specific visual problems of the client and on low vision in general. Carers were trained to get insight in visual problems by walking with blindfolds and spectacles to simulate visual impairment; how to give specific support embedded in daily routines (use of other senses, communication, stimulation of activities); spectacle habituation training and the importance of light, acoustics and contrast.

### **Outcome measures**

We followed the implementation process with a questionnaire for carers. Items used to measure aspects of familiarity, appreciation and implementation are shown in Appendix 1.

It would have been logical to use familiarity and appreciation scores at  $t_1$  for the analysis. However, not all questionnaires were completed at  $t_1$ , and in other cases, familiarity or appreciation scores were inconsistent during follow-up. Therefore, interpretation of the scores was done in the following way.

Appreciation scores were based on one item (see Appendix 1). To obtain one appreciation grade for each participant, appreciation was graded 2 if during follow-up 'yes' had been scored at least once and 'partially yes' never; the grade was 1 if 'partially yes' had been scored at least once and 0 if all scores had been 'no' or missing.



Scores for familiarity and implementation consisted of several items (see Appendix 1). A complicating factor was that advice could vary from just glasses, to a combination of ophthalmological treatment, specific support by carers, as well as technical improvement of the living environment. Therefore, objective familiarity and implementation grading was obtained through a panel. Four members of the research team received for each participant an overview of advised interventions and all item scores on  $t_1$ ,  $t_2$  and  $t_3$  (Appendix 1). Based on this information, total grades for familiarity and for implementation were assigned by each panel member independently, on a scale of 1 (worst) to 5 (best). Consensus was defined as one point difference or less between all panel members. All cases of disagreement were discussed, until consensus on a grade was reached. Grades assigned by the four panel members were averaged. This way each participant had one grade for familiarity and one grade for implementation.

### **Analysis**

Associations of familiarity, appreciation and implementation scores with severity of ID and of visual impairment were analyzed with Spearman's correlation. In the same way associations of implementation with familiarity and appreciation were analyzed. The level of significance was set at  $p < 0.05$ .

## **Results**

### **Study population**

Sixty adults (43 females; mean age 45 years; range 25-67 years) identified with a new diagnosis of low vision, blindness or subnormal vision that improved with at least 0,3 with glasses participated. Severity of ID was moderate (IQ 35 – 55) in twelve (20%), severe (IQ 25 – 35) in 38 (63%) and profound (IQ < 25) in ten participants (17%).

According to WHO criteria, 45 participants had a visual impairment and five were (socially) blind; visual function was subnormal in ten participants.

### **Advised interventions**

Glasses were prescribed for seventeen participants; two of them had inadequate glasses prior to the screening. In four of these cases, spectacle habituation advice was given. Forty-eight participants received written advice (Appendix 2a) on technical environmental adaptations and specific support by

carers aimed at communication and activities, conscious use of sounds, preferred places at the dining table and in the living room, routing, and better use of illumination day and night (electric light, sun screens). General advice to improve illumination, contrasts and acoustics was given in a majority of cases. However cheap measures were more frequently advised than expensive adaptations. The median time interval between screening and date of the written advice was 132 days (over four months). This delay was partly caused by the fact that first a report on the diagnosis was written and sent to the psychologist of the low vision center. This psychologist then made an appointment to observe the participant at home in daily activities before writing the advice.

### **Follow-up information**

The number of completed questionnaires decreased from 58/60 at  $t_1$  to 50 at  $t_2$  and 44 at  $t_3$ , resulting in three completed questionnaires with follow-up information on 43 and incomplete information on seventeen participants.

### **Familiarity, appreciation and implementation grades**

In 18/60 cases, disagreement about familiarity and/or implementation grades existed between panel members, but consensus was reached after discussion of the questionnaire scores. The mean grade for carers being familiar with the advice was  $3.69 \pm 1.22$  (median = 4) on a scale of 1 (completely unfamiliar) to 5 (completely familiar), with grades of 4.0 or more assigned to 41/60 participants. A positive appreciation grade (2) was assigned in 34 and a partially positive grade (1) in ten cases. The mean grade for implementation was  $3.95 \pm 1.25$  (median = 3) on a scale of 1 (nothing implemented) to 5 (completely implemented), with grades of 4.0 or more assigned to 20/60 participants.

Eight of the seventeen persons who received (new) spectacle prescriptions actually received spectacles and six were still wearing them at the end of follow-up. We do not know from our data why the prescribed glasses were not purchased or not used, but there is some anecdotal information. For example: parents considered their daughter “not a type for glasses”, therefore glasses were not purchased. Or the prescription was for reading glasses and the carer noted that the participant did not read and therefore did not need the glasses.

Familiarity, appreciation and implementation grades were not influenced by the severity of ID, but Spearman’s rho showed a small but significant positive correlation between severity of visual impairment and appreciation of the advice (Spearman’s rho 0.278, two-tailed  $p = 0.032$ ). Implementation was significantly

correlated with carers' familiarity with the advice (Spearman's rho 0.602, two-tailed  $p = 0.000$ ), but not with carers' appreciation of the advice (Spearman's rho 0.224, two-tailed  $p = 0.086$ ).

## Discussion

This is a first study on the quality of low vision treatment and rehabilitation for adults with intellectual disabilities (ID). We showed that advised interventions with glasses and in untreatable cases with specific support by carers and technical improvements in the living environment, are not always adequately realized in homes for people with ID. In no more than twenty out of sixty participants, the implementation was reasonable or complete, in spite of on-site information and training of the carer teams by experts of the low vision centers. Only six out of seventeen prescribed spectacles were still used after a year of follow-up. Implementation was significantly correlated with the familiarity of the carers with the advice, but not with their appreciation of the advice.

For a new and complex intervention, such a result might not be considered unfavorable, certainly considering the high turnover of carers. A positive finding is that we could not demonstrate a significant association of implementation with severity of ID. As far as we are aware no other implementation studies of treatment or rehabilitation for low vision, or specifically for refractive errors, have been published for this population.

We have recently presented evidence that low vision is associated with extra disability, on top of intellectual disability, in persons with moderate, severe and profound ID (Sjoukes et al., in press 1). This knowledge might convince carers of the necessity of intervention in this group. Unfortunately, well-designed studies into the benefits of interventions have not yet been published for the population with ID. In the current study individual examples were reported to us of improved social contacts, increased independence, explicit appreciation and use of better light, recognition of details, appreciation and use of brightly colored materials, uptake of new activities or old hobbies.

For technical adaptations, carers were dependent on allocation of budget by the management and on local technical procedures, which may have led to delays and with it, loss of awareness and motivation. Because of the very high risk of

visual impairment in the population with ID, a policy of obligatory optimization of illumination, colors and contrasts in all homes, day activity centers and workshops where people with ID live and work may be considered.

The procedures of the low vision centers may also have diminished the effectiveness of interventions, including the very long period between screening, where carers are first confronted with the limited visual functioning of their clients, and the receipt of advice reports (on average over 4 months), as well as the fact that concrete advice was hidden in long, academic reports (Appendix 2b). For carers, overloaded with practical tasks, such reports are discouraging. Moreover, technical advice often remained unspecified and limited to low-cost investments, possibly because of low expectations of the service provider's readiness to invest in existing buildings. Such an attitude may lead to sub optimal advice.

We conclude that even in high-income countries, many adults with intellectual disabilities and low vision lack adequate treatment and rehabilitation. The large numbers of unidentified low vision and blindness in the population with ID (Splunder et al., 2006) are of course an important factor. However, this study shows that even after on-site diagnosis, written personal advice, and specific training of staff, implementation within homes for adults with ID is fragmentary. Implementation of the advice is hampered by carers' limited knowledge of low vision followed by not understanding the written advice. From our questionnaire it became clear that once a carer is trained and educated on low vision and knows how to handle low vision problems of clients, this knowledge easily disappears because of the high turnover of caring staff. Therefore we think that sensory impairment should be an important subject in the education of carers.

The advising procedures can also be improved, we have recommended to Dutch low vision centers the following to improve effectiveness:

- Only screen when the board of the service provider agrees to support the implementation of advice;
- Reduce the time between screening and advice;
- Write the advice concise and easy to read;
- Start the advice with a summary of the most important points;
- Offer follow-up by telephone 3 – 6 months after the advice;
- Make sure that carers are (and stay) familiar with the advice by mentioning it in the yearly care plan;

- Make agreements that in case of (re)building facilities for people with ID the low vision center will advise on illumination and design in order to adjust buildings to the needs of people with ID and low vision;
- Because low vision centers and ID Service providers in most high-income countries do function globally the same, we do expect the situation in other countries to be similar.

### **Acknowledgements**

We thank the management and staff of the following Dutch intellectual disability services: Talant, Drachten; Amarant, Tilburg; ASVZ De Amerpoort, Baarn; and staff of the following low vision centers: De Brink, Vries; Sensis, Breda; Bartiméus, Doorn.



## Appendix 1

Information form used by research team to assign grades for appreciation; familiarity and implementation to each participant.

### • Appreciation grading

Question*		t <sub>1</sub>	t <sub>2</sub>	t <sub>3</sub>
Do you consider the advice practical?	yes/partially/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

### • Familiarity and implementation grading

Box below is colored red if advice has been given

Client number:

Glasses	Glasses habituation program	Treatment and communication	Illumination	Color
<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

## 1. Data which might influence the judgement on the implementation

Question*		t <sub>1</sub>	t <sub>2</sub>	t <sub>3</sub>
Questionnaire returned?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Did participant move home?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Did different persons fill out the questionnaires? (Each letter = name of a person, X = no name given)		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

## 2. To what extent is the advice familiar to the person who completed the questionnaire and to team members?

Question*		t <sub>1</sub>	t <sub>2</sub>	t <sub>3</sub>
Are you aware of an advice for the treatment of low vision?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Has the advice been discussed in the team?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
If yes, with how many team members?		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
The advice has been discussed with (cumulative t <sub>1+2+3</sub> )**		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Is the advice included in the personal care plan of participant?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Have you been trained on low vision?	yes/no/planned	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
How many team members were trained?		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
How many trained persons are still member of the team?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Do you know whether there is a prescription for glasses?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Did the participant get a prescription for glasses after the visual function assessment?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Is there a program on how to introduce spectacles to participant?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Are you aware of an advice on light?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Are you aware advice on the use of color and contrast?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Did you read the advice?	yes/partially/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

\* Empty box means: no answer received

\*\* The following persons were mentioned at least once: p = participant, t = tutor of participant, day = tutor in the day care center, d = doctor, ps = psychologist, f = family, TD = technical service provider, o = others

Indicate how well you think the team is familiar with the given advice.

**Totally unfamiliar** . . . . . **Completely familiar**

1	2	3	4	5
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### 3. Questions to assess the actual implementation

Question*		t <sub>1</sub>	t <sub>2</sub>	t <sub>3</sub>
Do you follow up the treatment advice?	yes/sometimes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
If no, why not?*				
Did you get ideas for an improvement for the participant?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Does the participant wear glasses?	yes/with certain activities/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
If no, why not?*				
Is the program to introduce glasses followed?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Has the light been improved as advised?	yes/partially/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Is the improved light used in the evening/ at night?	yes/sometimes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
If no, why not?*				
Is the improved lightening being used during the day?	yes/sometimes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
If no, why not?*				
Are walls, doors etc. painted in different colors as advised?	yes/partially/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Is better visible material purchased?	yes/no	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

\* Empty box means: no answer received

\*\* 1, 2, 3 followed by a reason, indicates the time the reason was mentioned

Indicate how well you consider the advice is implemented.

**Not implemented** . . . . . **Completely implemented**

1	2	3	4	5
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## Appendix 2

### Low vision rehabilitation advice

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At the outset of our study the rehabilitation advice provided by the low vision centers typically consisted of a five page professional document and as such was not always easy to understand for people involved in daily care. Examples of suitable, practical guidelines for carers are presented below in 2a. Examples of ‘difficult’ language that can best be avoided are presented in 2b.

#### 2a. Examples of good practical advice for a 44 year old woman with severe ID and low vision:

- “When you enter the room tell her your name and explain what you are going to do”
- “Tell her what is going on in the room, who else is there, etc.”
- “Slow down, she needs a lot of time to understand a request”
- “Use other than visual senses: tell her the coffee is being served and let her smell the coffee”
- “Give her a place in the living room such that she can see most with her right (best) eye”
- “Don’t change or move furniture without telling and showing her”
- “Avoid (background) noise and turn off the radio when you want to talk with her”
- “Keep the sunshades down to prevent glare”
- “Use contrast to make things better visible: use colored cups and plates on light colored unpatterned table(cloth)”

#### 2b. Examples of ‘difficult’ language

- The primary cause of her visual impairment is cerebral
- Beware that acoustic input is important
- Use contrast to make things better visible
- She has no problem with diffuse light
- Myopia is a refraction error of the eye
- Strong visual stimuli can improve her arousal
- Contrasting colors can be applied to facilitate orientation





**A randomized controlled trial  
of interventions for visual  
impairment in adults with  
intellectual disabilities**

Sjoukes L, Kooijman AC, Koot JM & Evenhuis HM

## Abstract

**Background** Our purpose in this multi-center randomized controlled trial was to investigate effects of treatment or rehabilitation of visual impairment in adults with intellectual disabilities during a follow-up of nine months.

**Method** Sixty cases and 45 controls with visual impairment completed the study. The diagnosis was made through ophthalmological assessments, applying WHO/IASSID definitions. Individual intervention advisories, given by experts of the low vision centers, included medical, environmental, professional and educational advice. Carer teams were trained on-site. Outcome measures were independent living skills, communication, locomotion, peculiar looking habits, self-absorbed and withdrawn behavior, antisocial and disruptive behavior, using three observant-based questionnaires. Multiple imputation was applied to control for missing values. The effect of the intervention was evaluated using the average change from baseline of each outcome during follow-up. This 'area under the curve' was compared with the 'area under the curve' of the control group using t-tests. All analyses were done on an intention-to-treat basis.

**Results** No significant differences were found between the intervention and the control group.

**Conclusions** We conclude that as yet, evidence supporting general screening of visual function in adults with intellectual disabilities, as has been recommended by an IASSID consensus group, is insufficient.

## Introduction

Adults with intellectual disabilities (ID) are an important risk group for visual impairment, with population-based prevalence rates over age 18 years of 13.8% (95% confidence interval (CI) 9.3 – 18.4%) for visual impairment and 5.0% (95% CI 3.8 – 6.2%) for blindness (Splunder et al., 2006). Unlike other frequent conditions in this population, such as motor impairments and epilepsy, this problem of visual impairment has long remained unrecognized. Up to this day, many persons with ID have to live with unidentified visual impairments (Splunder et al., 2006). Recently, we reported that, although intellectual disability itself has strong effects on daily functioning and behavior, visual impairment has significant extra effects (Sjoukes et al., in press 1). This concerns specifically peculiar looking habits, daily living skills, communication and language, and movement. Does this imply that intervention will result in improvement of daily functioning and communication? We designed a randomized controlled trial to answer the following question: what are effects of treatment and rehabilitation of visual impairment or blindness in adults with intellectual disabilities?

## Methods

### Study design

We set up this multi-center study as a randomized controlled trial (RCT). To realize this, we had to meet the following challenges:

1. Once visual impairment has been identified, it is unethical to delay treatment for research purposes. By consequence, randomization was performed before visual function assessment.
2. Because of the same ethical consideration, identification of visual impairment in those allocated to the control group had to be delayed until the end of the follow-up period. Normally, screening by Dutch low vision teams within care organizations for people with ID is performed on a monthly or bimonthly basis and it may take many months, even years, to complete all assessments. For this study, the participating low vision teams reserved considerably more time to complete all assessments within a limited time frame. Therefore, we considered a delay of assessments in the control group ethically justified.

3. With a population-based prevalence of around 20% visual impairment and blindness (Splunder et al., 2006), randomization before assessment would imply inclusion of a very large study population, of which a majority would appear to have (sub)normal vision. Therefore, we decided to select an 'at risk' group for informed consent, using an observant-based questionnaire.

The study was approved by the Medical Ethics Committee of the Erasmus University Medical Center (MEC 197.075/2000/239).

### **Study population and randomization**

Three Dutch service providers, together delivering ambulatory support, residential care, day care, and sheltered workshop services to around 5000 children and adults with ID, where a routine vision screening by a district low vision center had been scheduled for 2002 – 2004, consented to participate in the study. The three involved low vision centers adapted their screening to recommendations of the IASSID consensus guidelines (Evenhuis & Nagtzaam, 1998).

Clients aged eighteen years and over, with no previous diagnosis of visual impairment or blindness and no severe motor impairment or progressive disease (e.g. dementia or cancer) were eligible for the study. A power calculation was not performed, because of the absence of published data on effects of treatment and their size. We considered a group size of 100 in both trial arms practically feasible. Because of the population-based prevalence of 20% visual impairment and blindness, this would require recruitment of a base population of around 1000 persons fitting the inclusion criteria.

Our local contact persons asked the staff of homes and day activity centers to select clients fitting the inclusion criteria and complete an observation-based questionnaire to identify persons at risk for visual impairment. The questionnaire consists of 49 questions to be scored 'true' or 'not true': 14 questions on observable problems with the eyes (4 points per item if true), 16 questions on conspicuous behavior (1 point if true); 7 questions on reacting towards other people (2 points if true) and 12 questions on reacting towards objects (2 points if true). The questionnaire had not been validated, but staff of the low vision center that designed it, had the experience that 60% of those with a score of 4 points or over had a visual impairment. Written informed consent for participation in the study was asked from clients with a score of 4 points and over and/or their legal representatives.

Within each care organization, homes were randomly allocated to either the intervention or the control group on a 50 – 50 basis. Group size was not taken into account.

### **Outcome measures**

Social and behavioral functioning were assessed with three observant-based questionnaires in which the following five areas are addressed: independent living skills, communication, locomotion, peculiar looking habits, self-absorbed and withdrawn behavior, antisocial and disruptive behavior.

### **Measurement protocol**

The measurement protocol is generally outlined here. Detailed information on methods and definitions will be given in the paragraphs below.

Baseline measurements ( $t_0$ ) were performed in all persons who had consented to participate. Caregivers who knew the participants well completed the questionnaires. This was done prior to the screening of visual function, so caregivers were blind to the actual visual function of the participants.

Once the  $t_0$  measurement was taken, diagnostic assessments by the low vision teams started in the intervention group. After a diagnosis of visual impairment or blindness, individual advice for treatment or rehabilitation was given by experts of the low vision teams, and carer teams were informed and trained.

During a follow-up period of nine months, social and behavioral functioning were repeatedly assessed with the same three observant-based questionnaires as were used at baseline. This was done at three months ( $t_1$ ), six months ( $t_2$ ) and nine months ( $t_3$ ) after the intervention advice. For all participants in the control group, the same follow-up data were collected at three, six and nine months after the baseline measurement. To identify those with a visual impairment or blindness, diagnostic assessments in this group started after the follow-up period. The same procedure of advice and training was offered to the control group, but this was no longer part of the study.

### **Assessment of visual function**

Diagnostic assessments were performed by low vision experts on-site in the homes or day activity centers, and included at least two visual acuity tests (Snellen's chart, Burghardt's children's chart, STYCAR single characters and

matching, Cardiff Acuity Cards, Teller Acuity Cards), contrast sensitivity (Hiding Heidi), visual fields (confrontation), autorefraction or sciascopy, assessment of strabismus, handheld slit lamp biomicroscopy.

### Definitions and classifications

Information on the severity of ID was obtained from the client files, and was based on prior IQ testing. Severity was classified as mild (IQ 55 – 70), moderate (IQ 35 – 55), severe (IQ 25 – 35) or profound (IQ < 25). Visual function was defined as the visual acuity in the best eye (with optimal correction, if glasses were used). ‘Visual impairment’ and ‘(social) blindness’ were classified according to criteria of the World Health Organization (WHO) (1980). Clients who, according to WHO criteria, were not visually impaired but had subnormal distance or near visual acuity which improved 0.3 points or more with new spectacles, were also asked to participate. All participants were assigned to one of the ID and vision categories, presented in Table 6.1.

Table 6.1  
Classification of intellectual disability (ID) and visual function

ID level	IQ	Developmental age [years]	Visual function level	Visual acuity [arcmin <sup>-1</sup> ]
Mild	50 – 70	6 – 11	Normal vision	≥ 0.5
Moderate	35 – 50	3.5 – 6	Mild impairment	0.3 – 0.5
Severe	20 – 35	1.5 – 3.5	Visual impairment	0.05 – 0.3
Profound	< 20	< 1.5	(Social) blindness	< 0.05

### Intervention advice

Individual intervention advisories were prepared by experts of the low vision center including medical (glasses, ophthalmological treatment), environmental (improvement of light, color and contrast, routing), professional (training of staff), and educational advisories (information of clients and families). On-site training of the carer team involved explanation on specific outcomes and problems of the individual client and on low vision in general. Carers got insight



into visual problems by walking with blindfolds and spectacles specially prepared to simulate visual impairment. Training was given in specific support embedded in daily routines (use of other senses, communication, stimulation of activities), spectacle habituation procedures, and the importance of light, contrast and acoustics.

### **Behavioral questionnaires**

*Adaptive behavior scale (SRZ).* Adaptive behavior was measured with the Dutch 'Sociale Redzaamheidsschaal voor Zwakzinnigen' (SRZ) (Social Functioning Scale for people with ID) (Kraijer & Kema, 1994). The questionnaire consists of 31 items, each with four descriptions of possible behavior, from inapt to adequate, with corresponding rating values from 1 to 4. The item score is the rating value of the description that fits a participant's behavior best. The SRZ total score is the sum of all ratings and ranges from 31-124. Adaptive behavior is further specified with four subscales in which items are grouped: daily living skills (12 items), initiative & persistence (5 items), social skills (5 items) and communication & language development (9 items). The SRZ is found to be reliable and well validated (Kraijer & Kema, 1994). The correlation with the Vineland Social Maturity Scale is 0.82 (Kraijer, 2000). The instrument is widely used in the Netherlands for persons with moderate and severe ID aged 4 years and over.

*Vision Related Behavior Questionnaire (VRBQ).* The Vision Related Behavior Questionnaire (VRBQ) is a new questionnaire for the assessment of specific vision-related behavior in people with ID, that we developed because there was no suitable instrument available for the population of this study. An expert group consisting of behavioral scientists from the three participating low vision expertise centers was formed to define examples of vision-related behavior. The questionnaire consists of 45 behavioral items pertaining to problems with eye-hand coordination, eye-foot coordination, recognition of persons and objects, spatial orientation, mobility, anxiety, peculiar looking habits and communication. The items are scored in terms of occurrence: always, often, sometimes or never present, with corresponding rating values 1 to 4. A higher rating corresponds to more normal behavior.

Five clusters of vision-related behavior were identified using principal component analysis (based on 41/45 items with a factor loading  $\geq 0.4$ ): eye-hand coordination & mobility (9 items), recognition & communicative skills (9 items), eye-foot coordination (8 items), anxiety/ fear (6 items) and peculiar looking habits (9 items). The maximum score for each subscale is the number of constituent

items x 4. The VRBQ total score is the sum of all individual rating values and ranges from 45 to 180. The VRBQ has been shown to be reliable: Cronbach  $\alpha$  is 0.6383 (a scale is reliable when Cronbach's  $\alpha > 0.6$  (Malhotra, 1993)).

*Development Behavior Checklist (DBC).* The Development Behavior Checklist (DBC) was developed to identify emotional and behavioral problems in children with ID (Einfeld & Tonge, 1995). It has been translated and validated for Dutch children, but it proved to be applicable to adults as well (Dekker et al., 2002; Ruiter et al., 2007). The DBC includes 95 descriptions of problem behavior that are scored as either not true, partly true, or true, with the rating values 0, 1 and 2. Higher scores indicate more emotional and behavioral problems. The DBC has the following five, partly overlapping subscales: disruptive & anti-social behavior (27 items), self-absorbed behavior (31 items), communication disturbance (13 items), anxiety (9 items), and social relating (10 items).

### Analysis

Missing values were multiply imputed using chained equations (Buuren et al., 1996). Each missing value was replaced with five different estimates, thus taking the uncertainty of the imputation into account. The imputed data sets were analyzed separately and later combined using 'Rubin's rules' (Rubin & Schenker, 1991). Characteristics of the intervention and the control group were compared with the student's t-test to find out if there were significant differences in sex, age, level of ID or level of visual impairment. Total and subscale scores of the three questionnaires at  $t_1$ ,  $t_2$  and  $t_3$  were computed. We used a linear mixed model to model the various outcomes as a function of time. All analyses were controlled for levels of visual function and ID. To take account of the design of the experiment, the individual and group were included as random effects. Furthermore, we allowed auto correlation and heteroskedasticity in the residual covariance matrix. The effect of the intervention was evaluated using the average change from baseline of the outcome. This 'area under the curve' was compared with the 'area under the curve' of the control group using a t-test (with the Satterthwaite approximation for degrees of freedom). All analyses were done on an intention-to-treat basis. Results were considered statistically significant when the two-tailed p-value was less than 0.05. Multiple imputation was done using IVE ware, the mixed model was estimated in SAS 9.1 and all other analyses were done using SPSS 15.0.

## Results

### Study population

Inclusion, randomization and participation are presented in a CONSORT flowchart (Figure 6.1). In 140 homes or day activity groups, carers selected 697 clients fitting the inclusion criteria. After pre-screening, informed consent was obtained for 292/340 persons (86%); no consent was obtained in sixteen cases (5%), whereas 32 clients (9%) did not react. Sixty cases and 45 controls completed the study.

Characteristics of the intervention and control groups are shown in Table 6.2. The student's t-test showed no significant differences of sex, age, level of ID or level of visual impairment.

Table 6.2  
Characteristics of study population (%)

Characteristics	Intervention group N = 61	Control group N = 45
Male	29.5	37.8
Mean age [yrs] (range)	45 (25-67)	46 (23-74)
Mild ID	-	2.2
Moderate ID	19.7	40.0
Severe ID	62.3	42.2
Profound ID	18.0	15.5
Low vision	75.4	80.0
(Socially) blind	8.2	2.2
Visual acuity improved $\geq 0,3$ with glasses	16.4	17.8

### Interventions

Glasses were prescribed for seventeen participants; two of them had inadequate glasses prior to the screening. In four of these cases, spectacle habituation advice was given. Forty-eight participants received written advisories

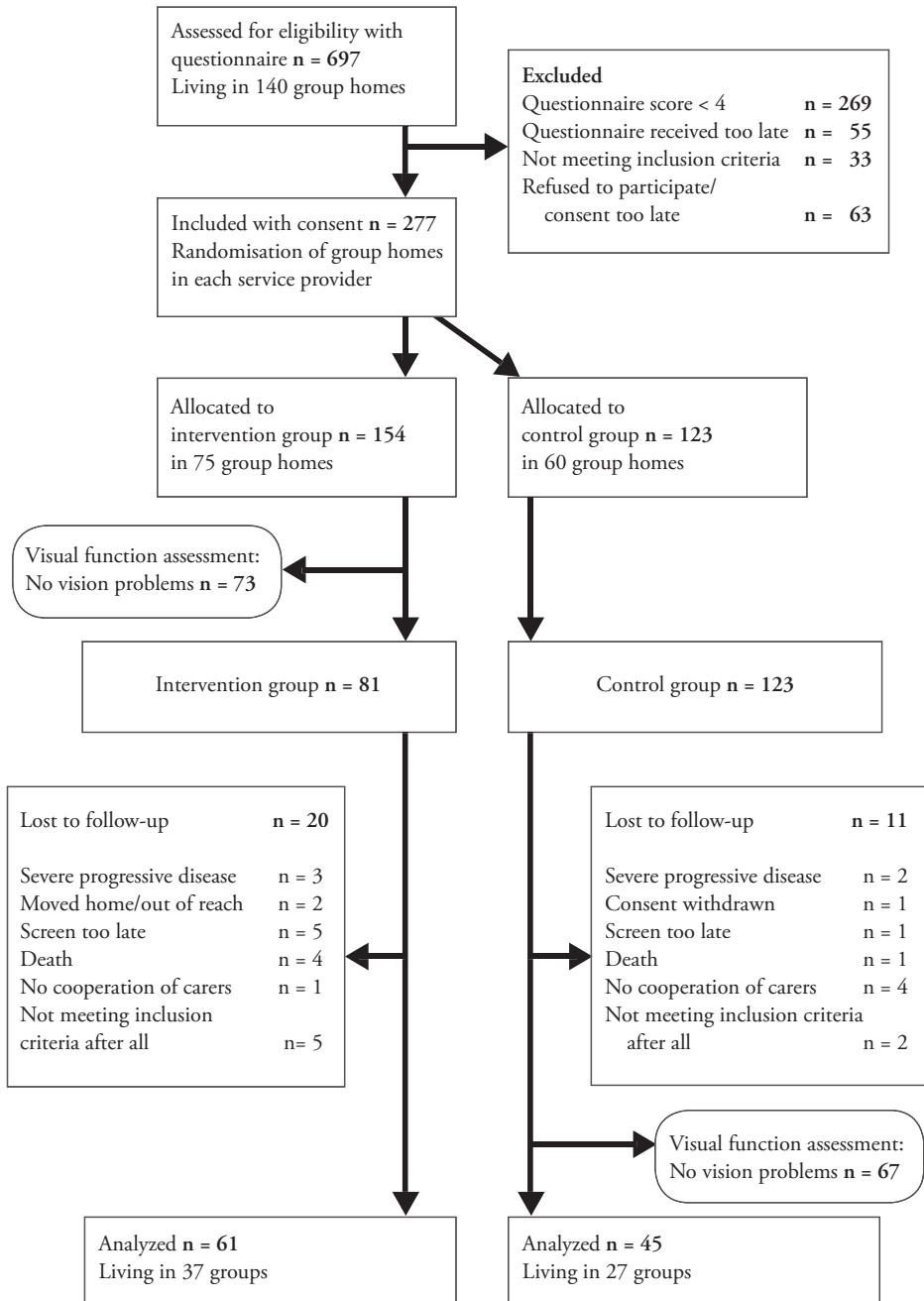


Figure 6.1 Visual function assessment: flowchart of inclusion, allocation and participation

on technical environmental adaptations and specific support by carers aimed at communication and activities, conscious use of sounds, preferred places at the dining table and in the living room, routing, and better use of illumination day and night (electric light, sun screens). General advice to improve illumination, contrasts and acoustics was given in a majority of cases. During follow-up, a reasonable to complete implementation of interventions was reached in twenty out of sixty experimental cases. This was not correlated with severity of ID (Sjoukes et al., in press 2).

### **Follow-up information**

On average, questionnaires were returned after four ( $t_1$ ), eight ( $t_2$ ) and twelve months ( $t_3$ ), extending length of follow-up to twelve months. The number of completed questionnaires for the intervention group decreased from 58 at  $t_1$  to 50 at  $t_2$  and 44 at  $t_3$ , resulting in complete follow-up information on 43 and incomplete information on 17 participants. For the control group, 45 questionnaires were completed at  $t_1$ , 44 at  $t_2$  and 35 at  $t_3$ .

### **Outcomes**

Table 6.3 shows that no significant differences between the intervention and the control group were found in the 'area under the curve' of SRZ, VRBQ and DBC total scores. The same applies to all subscale scores (data not shown).

## **Discussion**

Although we have demonstrated in an earlier study, that visual impairment and blindness are associated with significant extra disability in adults with intellectual disabilities (ID) (Sjoukes et al., in press 1), this randomized controlled trial could not demonstrate any significant effects of treatment and rehabilitation on functioning and behavior during one year of follow-up.

This outcome does not support the evidence for screening as advised in the IASSID consensus statement on identification of visual impairment (Evenhuis & Nagtzaam, 1998). However, this is not the moment to jump to the conclusion, that intervention for visual impairment in this population is useless. In spite of the careful preparation of the trial, the negative result can be partly explained by methodological issues, such as sample size slippages, incomplete implementation of interventions, and incomplete observations of outcome measures by carers.

Table 6.3  
Average change from baseline ('area under the curve') for the intervention and control group, and their differences in the total scores of each questionnaire

Group	Questionnaire	Estimate	Standard error	95% confidence interval	p-value
<b>Intervention</b>					
	SRZ total	-0,477	1,467	-3,545–2,591	0,749
	VRBQ total	-1,388	1,410	-4,211–1,436	0,329
	DBC total	-2,521	2,256	-7,070–2,028	0,270
<b>Control</b>					
	SRZ total	2,379	2,145	-2,527–7,286	0,298
	VRBQ total	0,032	1,652	-3,287–3,351	0,985
	DBC total	3,707	2,589	-1,506–8,919	0,159
<b>Difference</b>					
	SRZ total	-2,856	2,232	-7,516–1,804	0,216
	VRBQ total	-1,420	2,085	-5,571–2,731	0,498
	DBC total	-6,228	3,223	-12,643–0,187	0,057

Implementation was far from complete, in spite of involvement of management and staff in coordinated preparations (Sjoukes et al., 2006; Sjoukes et al., in press 2). High turnover and workload of staff may have played a role in knowledge and motivation leaking away during follow-up. Similar findings have been reported in an RCT of bereavement support by carers (Dowling et al., 2006). The authors observed that the intervention appeared often to be given a low priority.

During recent years, the feasibility of the RCT design in study populations with ID has been discussed by several authors. Fidelity or incomplete implementation of complex interventions may compromise trial outcomes in persons with intellectual disabilities (Guralnick et al., 2006; Bagner & Eyberg, 2007). Voluntary participation based on self-referral led to better motivation of parents to complete the study (Plant & Sanders, 2007), but may lead to bias in

the study population. Regular supportive face-to-face contact with the researchers may also stimulate motivation (Montgomery et al., 2007). Recruitment is generally recognized as difficult because of barriers with service providers (lack of resources, workload of carers, lack of familiarity with research, organizational barriers) and problematic informed consent (Oliver et al., 2002; 2005; Martin et al., 2005; Kerr et al., 2005; Lennox et al., 2007). In our study, recruitment was problematic only at the level of service providers, as we had to approach 15 large service providers to find three organizations prepared to participate. This is disappointing in the light of the high consent rate of 86%. Because of the limited size of the population with intellectual disabilities, to obtain a large enough study population, inclusion criteria are usually kept as unrestricted as possible. This resulted in heterogeneity of the study population in published studies as well as the current study, influencing both interventions and outcomes (Oliver et al., 2002; 2005; Martin et al., 2005; Kerr et al., 2005; Hellings et al., 2006; Montgomery et al., 2004).

In our study, the outcomes might have been more complete if the questionnaires had been completed face-to-face with the researcher. On the other hand, observant-based questionnaires may not be the best instruments to monitor small behavioral changes. Video images might more sensitively register such effects, but interpretation is very time-consuming.

We conclude that further research into effects of treatment and rehabilitation of visual impairment, preferably in children and young adults, is required before starting new screening programs. To start with, participants with mild and moderate ID may be more easy to motivate and evaluate. Systematic evaluation of the attainment of personally formulated goals might be a model to ensure cooperation and implementation.

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# 7

## **Inadequacies in the treatment of visual impairment in people with an intellectual disability: Who is responsible?**

Evenhuis HM & Sjoukes L

Heleen Evenhuis and Liesbeth Sjoukes were asked to write a comment on the findings published by van Isterdael et al. (2008) in the Dutch Medical Journal on a retrospective file study of the past decade of referral of people with intellectual disabilities (n = 5.205) to a third line expertise centre for low vision or blindness.

## Comment

Van Isterdael et al. (2008) ask attention for the treatment of refractive errors and cataract for people with intellectual disabilities. This is important because data on ophthalmological treatment in this population are hardly available, certainly not in such a large population. The analysis is based on files of Bartiméus, a service for people with visual impairment, which was the first to start screening and offer advice to service providers for people with intellectual disabilities.

Coworkers participated in the development of a multidisciplinary consensus guideline to diagnose visual problems in this population. (Nagtzaam & Evenhuis, 1999). Bartiméus and sister organization Sensus have published a book on the design of the living environment for visually disabled persons, with a technical supplement for architects.

### **Visual problems of people with intellectual disability**

People with intellectual disability are at high risk for visual problems. Low vision and blindness in the Dutch population of people with intellectual disabilities is ten times more prevalent as in the general population of people aged 55 and over.

No data are available on the risk for children. The prevalence must be higher than in the general population because in populations of children with severe low vision or blindness the majority also has an intellectual impairment (Mervis, Boyle & Yeargin-Allsopp, 2002). The Netherlands therefore included, probably as the only country in the world, the population with intellectual disabilities as a target group in the WHO-project 'Vision 20-20' (Limburg, 2005). This project is focused on banishing avoidable low vision and blindness.

### **Consequences for functioning**

Many people doubt whether a visual problem leads to additional functional impairment for people with an intellectual disability and therefore are not convinced of the necessity of screening. In a recent cross sectional study we showed that screening does make sense (Sjoukes et al., in press).

Low vision leads to important additional impairment, especially in the activities of daily life, in communication and in mobility. It is plausible that timely treatment and rehabilitation of low vision will diminish such impairments.

### **Often treatable causes**

Van Splunder (2004) has mapped the treatable causes of low vision. Refractive errors are just as prevalent as in the general population, about 60%, but they are often more severe: the risk of severe myopia was 3.5 times higher. Of those needing glasses 52% had no or no adequate glasses. People with Down syndrome develop cataract about thirty years earlier than in the general population. For people with intellectual disabilities of other causes the prevalence of cataract increases about ten years earlier. This may be caused by pre-existent syndrome related cataract and by auto mutilation.

### **Inadequate treatment of refractive errors and cataract**

Van Isterdael et al. evaluated the treatment of refractive errors and cataract in this population. Their retrospective study involved more than 5.000 persons with intellectual disabilities, living in specialized institutions. Over a period of ten years they were referred by physicians for intellectual disabilities to third-line medical care for their visual functioning. Thus it was a selected population with an overrepresentation of persons with severe intellectual disabilities (66%). Follow-up data were only available for a minority; therefore the analysis was mainly on the given advice and for a small part on the implementation of the advice. Of those with a new diagnosis of a refractive error, only 28% received a prescription for glasses. Of all patients of 45 years or older, for whom presbyopia can be expected, no reading glasses were prescribed in 78% of the cases, even though these people can profit of such glasses when working on their day activity center, looking at photo's, jig sawing, knitting or doing needlework. Glasses were not prescribed if patients had severe intellectual disabilities or the diagnosis 'neurological visual pathology'.

Follow-up data were available for part of those with a prescription of glasses: the glasses were used in 84% of the cases. Of those with cataract (defined as any visible cloudiness of the lens and a visual acuity < 0.3) 45% was referred to an ophthalmologist; of those referred only 26% underwent surgical treatment. The authors conclude that apparently treatable conditions are left untreated in many cases.

### **Weaknesses of the study**

The disadvantage of retrospective file studies is that the researchers only have the file data. Therefore the study raises many questions, besides the importance of the given information, as the authors do acknowledge themselves.

- For how many persons glasses or cataract surgery were not considered indicated, because of multiple pathology, behavioral problems, dementia, or because of low expectations of improvement or acceptance by the patient?
- Which parties were mainly responsible for the decision on ophthalmological referral and treatment: the patient, the family, the professional carers, physicians for intellectual disability, general practitioners, ophthalmologists?
- Which considerations lead to the decision not to follow the advice? Why had people been referred while treatment apparently was not considered necessary in advance?
- And not unimportant: specialized third line diagnosis and advice is paid for by tax money in the Netherlands, it is time consuming and expensive. Are we too careless in handling the results? And are not mainly the care providers for people with intellectual disabilities responsible?

### **Implementation of treatment and rehabilitation**

In our own prospective study (Sjoukes et al., in press) on the implementation of advised interventions we experienced the same problems as Isterdael et al. (2008) The advice for sixty adults with a recent diagnosis of low vision or blindness were heterogeneous and could consist of a combination of one or more of the following: glasses, ophthalmological treatment, specific support in daily life and/ or technical improvement of the living environment (more light, sunscreens). The carer teams were trained by low vision expertise centers, while agreements were made with the care providers on organizational and financial conditions. Nevertheless in the 12 month follow-up period advice was reasonably implemented in only twenty of sixty cases. Of the seventeen prescribed glasses eight had been purchased and after 12 months six were still used. The implementation was not worse for people with more severe intellectual disabilities.

In 2005 we did a survey on ten randomly chosen boards of directors of service providers. All knew of the increased risk of visual (and hearing) impairment of their clients and of the large numbers of undiagnosed problems. All care providers had made deals with low vision expertise centers for free screening and advice. Most of the interviewed managers did not know the results of these screens, and did not know what happened with the advice. As possible factors for this low priority of treatment and rehabilitation they mentioned the lack of external pressure by family and government rules, and the fact that low vision does not lead to problem behavior for the professional carers.

The study of Van Isterdael (2008) leaves many questions unanswered. Based on our own study results we conclude that treatment and rehabilitation of low vision in the care for people with intellectual disabilities is hampered by lack of organizational and financial support, insufficient attention for visual impairment in the training for nurses and carers, high workload, frequent changes in staff, vacancies and insufficient pressure by insurance companies and government.

#### **Which physicians should feel responsible?**

The intellectual disability physicians and general practitioners can be blamed for the low referral practice. They should, together with the patient, family and other carers, actively advocate the importance of treatment keeping the severity of the intellectual disability in mind. During the epidemiologic research by van Splunder only few of the responsible general practitioners followed the referral advice without urgent request of the researcher and this concerned mostly people with mild intellectual disabilities. We recommend research into the considerations of ophthalmologists to decide pro or against cataract surgery and the development of guidelines for cataract surgery in this group based on effect research.

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# 8

## General discussion



This complicated study, designed as a randomized controlled trial, provided important insights, both scientific and on barriers to deliver quality of care. We were the first to demonstrate that visual impairment significantly hampers daily functioning, communication and mobility in adults, in addition to functional limitations as a result of the intellectual disability. However, treatment and rehabilitation are problematic on different levels: client, family, staff of homes, service providers for people with intellectual disabilities and low vision teams. As a result, implementation of treatment and rehabilitation advice was fragmentary and we were not able to demonstrate effects of interventions.

### **Additional disabilities by visual impairment and blindness**

One of the factors that may hamper diagnosis and treatment of low vision and blindness is, that it is literally and metaphorically an “invisible” handicap. We have demonstrated significant functional and behavioral differences between adults with and without visual impairment (Chapter 2). Generally, in children, visual impairment and especially blindness are associated with retardation in motor development (Thompson, 2002), attachment and learning skills (Levtzion-Korach, 2000; Dale, 2002; Atkinson, 2002). We suppose that if visual impairment is diagnosed and treated as early in life as possible, the general development might benefit both in children with normal intelligence and in children with a developmental delay. Part of the visual impairment in our adult study population may have been age-related visual loss. Sørensen (1995) has shown that simply increasing illumination in homes of elderly people in Sweden to adequate levels has clear, long lasting positive effects such as less depressive symptoms, better social contacts, less feeling of loneliness, better general health. Based on clinical practice, we expected the following effects of treatment or rehabilitation: improved mobility, better mood, and better performance on activities of daily life.

### **Effects of treatment and rehabilitation**

A systematic review (Chapter 3) of the available scientific literature, however, revealed that, apart from case reports or small case series, hardly any trials have been performed on effects of treatment or rehabilitation in groups with intellectual disabilities or related conditions (cerebral palsy, prematurity). The methodological quality of the few published trials is mostly poor: small and selected study groups; no controlled designs. Outcome measures concerning activities of daily life or behavioral aspects are lacking or non-validated. On the other hand, we found that knowledge on the effectivity of treatment and rehabilitation of visual impairment and ophthalmological conditions in the

general population is also primarily based on clinical experience, case studies, ophthalmological file studies and small, uncontrolled or non-randomized trials. Nevertheless, current methods of treatment and rehabilitation are generally accepted.

We conducted a randomized controlled trial in an adult population with intellectual disabilities (Chapter 6). However, in spite of careful preparations, both organizational and methodological, we were not able to demonstrate any effects of treatment or rehabilitation. Sample size slippages, incomplete implementation of interventions, and incomplete observations of outcome measures by carers were the primary methodological flaws. As a result, we cannot yet conclude that intervention for visual impairment in this population is useful.

### **Implementation is a major problem**

We anticipated that the implementation of this complex intervention would be a major obstacle (Oliver et al., 2002). We used concept mapping in multidisciplinary groups in the participating care organizations to facilitate and optimize the first steps of the implementation process. We aimed at: 1) increased awareness of low vision in professionals involved in treatment and care of people with intellectual disabilities, 2) identification by staff of possible barriers to implementation of treatment and rehabilitation (Clemmer, 1998), and 3) suggestions by staff of possible solutions (Chapter 4). The list of obstacles was brought to the attention of the boards of directors to obtain optimal support and means for the interventions.

Despite these efforts, the implementation of advised treatment was not adequately realized in about 2/3 of the cases (Chapter 5). We conclude that identification of anticipated obstacles is not enough to implement innovations without real obligations and engagement of the management. In this case, with a complex intervention requiring cooperation and time of many different carers and professional staff, expert project management including support, financial budget, embedding in routine procedures and well-designed scenarios would have been necessary to obtain better and lasting results.

From the literature two ways are proven effective to enforce implementation. Grol (1992) has shown that (financial) incentives may help to stimulate implementation. Another way to enforce implementation is by formal regulations. The Netherlands Board for Health care Institutions (College Bouw Zorginstellingen, 2002) has published technical recommendations for the living environment of people with visual and hearing impairments. For visual

impairment, these recommendations were based on expertise of the low vision centers (Wildenberg et al., 2002). A telephone enquiry in a random sample of service providers for people with intellectual disabilities made clear that since our research project the top management of care providers is more aware of the importance of an optimal living environment for people with low vision (Wiersema & Evenhuis, 2005). Therefore in case of renovation or new buildings, the low vision centers are regularly asked for technical advice. In our opinion, optimal conditions for low vision, like optimal acoustics (Meuwese-Jongejugd et al., 2005), should become compulsory in all buildings of intellectual disability services.

### **Is there evidence to support screening?**

We now address an underlying question, leading: is there now a sound scientific evidence to support screening for low vision in adults with intellectual disability, as recommended in Dutch and IASSID consensus statements (Nagtzaam & Evenhuis, 1999; Evenhuis & Nagtzaam, 1998)? According Wilson (1968) the criteria shown in Table 8.1 should be met.

Table 8.1  
Wilson's criteria for screening

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The disease	– should be an important problem;
	– there must be a recognized latent of early symptomatic stage;
	– the natural history must be understood.
The screen	– a suitable test or examination must be available;
	– the test must be acceptable to the population screened;
	– screening must be a continuous process.
Follow-up	– facilities must exist for assessment and treatment;
	– there must be an accepted form of effective treatment;
	– there should be an agreed policy on whom to treat.
Economy	– the cost must be economically balanced in relation to possible expenditure on medical care as a whole.

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The criteria for 'the disease' have now all been met, as are the first two requirements of 'the screen'. However, screening is currently not a continuous process in the Netherlands: most service providers delegate this to low vision teams and continuity is not guaranteed.

Moreover, usually no screening is offered to clients using community-based care, including many older persons and persons with Down syndrome. Recently, restrictions in financial compensation through the modified chronic health care insurance (AWBZ) have led to discussion in low vision organizations, to what extent screening should remain a task for tertiary specialist care. An e-mail inquiry among members of the Dutch association of intellectual disability physicians (NVAVG) showed that the majority considered screening to be problematic, either for practical reasons or because population screening is not considered a task of individual physicians (Walinga & Graaf, 2006). At present we are investigating the feasibility of screening by optometrists in local shops and ophthalmology departments, finding the Dutch association of optometrists (NVO) interested to participate.

A recent feasibility pilot with 14 residents of two Rotterdam community-based homes, performed in two shops, has shown that optometric assessment in this group requires more empathy, patience, flexibility and time compared to persons of normal intelligence, and therefore requires experienced optometrists. A familiar carer has to be present for support and information. Lacking or incomplete file information hampers efficient assessment (unpublished data). Most participants could be assessed reasonably well or good. Reimbursement by AWBZ or insurance companies is still a barrier to be taken.

This thesis and the study by Isterdael et al. (2008) (Chapter 7) both show that Wilson's criteria for 'follow up' are not being met in the Netherlands. Although facilities for assessment and treatment, in this case ophthalmologists and low vision teams, are available everywhere in this country, referral by intellectual disability physicians and general practitioners is unsatisfactory. Although in clinical practice clients clearly benefit from spectacles, cataract surgery, or optimal illumination, treatment and rehabilitation are evidently not a priority of service providers and are sometimes insufficiently accepted by family and staff of homes. Moreover, we could not validly investigate effects of intervention.

We conclude that screening of the adult population with intellectual disabilities is insufficiently evidence-based. The results of this thesis led to

discussions in the expert group on how to update the 1997 Dutch consensus guideline. Screening of subgroups and a different approach of groups with mild and moderate versus with severe and profound intellectual disabilities are now being considered. For children at risk for low vision, i.e. children with intellectual disabilities and/or cerebral palsy and prematurely born children, a detection model through routine youth health care has been developed in 2006 by a multidisciplinary group of pediatricians, pediatric ophthalmologists, intellectual disability physicians, youth health physicians and orthoptists (Evenhuis et al., 2007). First evaluation in a test region is in progress.

### **Considerations for intellectual disability care**

For each study in the field of intellectual disability cooperation is needed from legal representatives and staff to get consent for assessments and treatments as well as participation in research. They judge the benefits and burdens of interventions and participation in a research project. Considerations may be emotional and not always rational ('my daughter is not the type for spectacles'), which for family is understandable, but should not be so for staff. In our experience, staff in long-term care tends to be more concerned about expected burdens for their clients than about possible benefits. Fortunately, within the boards of Dutch service providers, the insight is increasing that development of new knowledge and scientifically evaluated care are relevant. Research should become more usual in this field and when positive results are obtained, the benefits will become clear.

### **Quality of advice by low vision expertise centers**

The low vision expertise centers, responsible for information, training and intervention advice, were surprised by the finding of the low implementation grades. This thesis shows that behavioral experts gave insufficiently specified technical advice to improve the living environment in a majority of cases. The advised technical improvements were not or only partially realized by intellectual disability service providers, probably because of financial or procedural obstacles. The low vision organizations planned to change their procedures to improve acceptance and implementation of their advice. From the concept mapping sessions they learned what kind of questions should be answered to motivate caregivers to follow their advice. Language and readability of advice and oral clarification to clients, teams of caregivers and family were improved. Indeed, Kahan (1988) analyzing implementation of American guidelines, found that those with emphasis on practical and clear recommendations on how to act were more likely to be followed than those with long theoretical descriptions. The top

management of one of the low vision organizations started an extensive innovative project to improve implementation. The project was set up with strict agreements with the management of intellectual disability service providers, newly designed educational materials, short and to-the-point written advice, joint formulation of individual aims of treatment or rehabilitation of carers and low vision experts and systematic individual evaluation by means of goal attainment scaling. This can be seen as an important and practical direct spin off from our research project.

### **Considerations for further research**

Follow-up of our research is necessary to find out if treatment and rehabilitation for people with ID with glasses, ophthalmic care and improvement in lighting, color and contrast of the environment have beneficial effects. For people who themselves cannot express beneficial effects other ways must be found to objectively assess effects. We used observational questionnaires but video observations or personal observations of specialized researchers may provide more specific and detailed information.

There is strong evidence that the medical treatment of visual problems of people with ID is not comparable with the care in the general population in the Netherlands. (Isterdael, 2008). Therefore a prospective follow-up study on referral of people with ID to an ophthalmologist compared to the referral procedure in the general population is quite necessary.

For people with ID with severe refraction errors spectacles sometimes are insufficient and contact lenses too difficult to use. For this group new techniques are promising: laser assisted keratoplasty is possible under full anaesthesia for children with severe myopia or hypermetropia who could not be treated with glasses or lenses. The results are very promising and the treatment seems to be viable for people with ID. A pilot study to investigate the benefits and side effects should be undertaken as soon as possible with careful follow-up.

Another research area is the treatment of keratoconus, a disease which is more common in people with ID especially Down syndrome. From the literature it appears that keratoconus is often only treated surgically after periods of severe hydrops of the cornea. In general keratoconus is treated with keratoplastic surgery in an earlier stage, before hydrops occurs, which probably explains to some extent the better results compared to those in Down syndrome (Völker-Dieben, 1993). Timely intervention may lead to better results for people with Down syndrome. Further research is necessary.



### **Conclusion**

A main finding of this thesis is that when intellectual disability co-occurs with visual impairment, the overall disability is more severe. Therefore treatment is indicated and since there is almost no evidence against usual treatment and rehabilitation for visual problems in people with ID, all usual treatments should be offered, because people with ID deserve the same level of health care as the general population.





## Summary



This thesis describes the results of a study that is a sequel to the results of an earlier study by J. van Splunder.

Van Splunder studied a random sample of 1.600 persons from a population of 9.000 with intellectual disabilities. According to the criteria for visual impairment and blindness of the World Health Organization approximately 15% of the sample had visual impairment and 5% was blind. Of the elderly with Down syndrome and severe intellectual disability even four out of five had visual impairment or was blind. Another important finding of Van Splunder was that in 40% of the cases caregivers and professionals were not aware of the visual impairment or blindness. He concluded that people with intellectual disabilities receive insufficient ophthalmological care.

Heleen Evenhuis, professor in intellectual disability medicine at Erasmus MC, prepared a study that should focus on the effect of treatment for adults with intellectual disabilities, once the visual impairment or blindness was diagnosed. This thesis describes the design, the realization and the results of this study.

First the participants for the study had to be found: adults with intellectual disabilities. This is possible through service providers for this population offering residential homes, sheltered workshops and day care. It appeared to be difficult to convince the management of the benefits of participation. Of fifteen service providers only three agreed to participate in this study and support and implement the treatment advice for clients who were diagnosed with visual problems.

Some people think that an additional problem like visual impairment does not make much difference for people with intellectual disabilities because they already are disabled. **Chapter 2** describes the study question whether people who are visually impaired have more problems with activities of daily life than people with normal vision. In order to assess this, measuring instruments had to be found. Observational questionnaires existed for people with intellectual disabilities. Other questionnaires existed for people with visual impairment, but none were found for people with both impairments. It was decided to use two well known and valid questionnaires: the Sociale Redzaamheidsschaal Zwakzinnigen (SRZ) by Kraijer (1994) and the Development Behavior Checklist (DBC) by Einfeld and Tonge (1995).

The SRZ measures the level of functioning in activities of daily life – taking a shower, getting dressed, setting the table and the like – for people with intellectual disabilities. The DBC is developed to measure emotional and behavioral problems in children with intellectual disabilities and is also used for adults. Neither of these questionnaires refers to specific visual behavior like eye hand coordination or visual recognition. For that reason a third questionnaire was developed to gather data on visual behavior. This questionnaire was constructed together with staff of the low vision centers involved in this research project: Sensis, Bartiméus, De Brink and Visio.

The three questionnaires were completed by caretakers for 269 participants. Afterwards the visual function was measured. Of the 269 persons 107 were diagnosed with low vision or blindness and they scored significantly lower on the ability to do things independently. Their eye hand coordination and mobility was significantly lower than in people with normal vision. It is shown that low vision indeed causes additional impairment in daily life in a clinically relevant way.

**Chapter 3** reviews the literature on treatment of eye problems and low vision in people with intellectual disabilities. Many case reports are written on this subject, but controlled studies on treatment results, especially on functional effects, hardly exist. On the other hand, most studies report that people with intellectual disabilities can be treated as usual, with some extra care in the pre and post operative period if needed. The conclusion of the review is that prescribing and wearing glasses, cataract extraction, occlusion for amblyopia or surgery for squint are feasible for people with intellectual disability in the same way as in the general population. Future research should include follow-up studies to objectively measure the results. New techniques like laser assisted eye surgery are being used on a small scale in this population, especially for people with severe intellectual disabilities who could not wear glasses or lenses. In those cases good results are reported.

Low vision in people with intellectual disabilities often remains untreated. Therefore it was of ultimate importance in this study to make sure that the recommended treatment was actually given. **Chapter 4** describes how all people involved in any aspect of treatment or rehabilitation were informed. A treatment advice can be complex. Close cooperation of caretakers, managers, psychologists, physicians and technicians is necessary to implement such complex advice. The advisory staff of low vision centers also plays an important role. In each participating institution all these parties were invited together for two sessions to

list all problem areas and obstacles that could be foreseen. A detailed overview was made with concept mapping techniques and this was brought to the attention of the management board. Agreements were made on how to solve problems (if possible) and handle obstacles.

After this extensive preparation sixty persons were diagnosed with low vision. Each of them received a treatment advice. **Chapter 5** describes how caretakers evaluated the advice and to what extent it was followed. In a twelve month follow-up period the caretakers completed a questionnaire three times. They answered questions like: Do you know whether an advice is given? If yes, do you know what the advice is about? Do you think the advice makes sense? Do you carry out the advice? If no, why not? The answers showed that in 2/3 of the cases the advice given was known to the caretakers and more than half of them considered the advice as useful. In only 1/3 of the cases the advice was carried out to a reasonable degree. The adherence to the advice was not influenced by the severity of the intellectual disability.

**Chapter 6** shows the results of the core research question: What is the effect of treatment? In the study a treatment group was compared with a control group. Participants were randomly assigned to either group: a randomized controlled trial. This type of research is not yet done very often in the population with intellectual disabilities. It yields the most valid results but is often very laborious and sometimes involves ethical problems. In this study the problem was that once the diagnosis was made it was unethical to postpone treatment for the sake of the study. That is why vision screenings were used that had already started in the three participating organizations. These screenings took place on one day once a month and it would take several years before all clients were screened. In the research year of this study it was agreed with the low vision centers to screen more people in the beginning of the year, than make a pause and again screen more people at the end of the year. In this way the same number of people or even more could be screened in one year and on the other hand those on the waiting list could be assigned to the control group. The control group was diagnosed at the end of the year and received advice after our study if necessary. All questionnaires were completed at the same time for both intervention and control groups. No effects could be shown in the treatment group. We cannot conclude however that treatment does not make sense, because it was often inadequate.

**Chapter 7** comments on a Dutch research project on people with intellectual disabilities who are referred to an ophthalmologist for refractive errors

and cataract. The retrospective file study shows that after referral treatment is poor. The comment focuses on the question as to who is responsible for good care in case of visual impairment. By now the management of service providers for this population has become more aware of the large numbers of visually impaired people who do not receive necessary treatment. A complicating factor is that visual impairment is 'invisible': the patient does not complain, does not seem to be unhappy and does not show challenging behavior. Therefore people in the daily environment of the person with low vision do not notice any problem. Even relatives are not always motivated for treatment as is shown by the following remark: "My daughter is not the type for glasses". It is concluded that the physician for people with intellectual disability or the general practitioner can estimate the advantages and the disadvantages of treatment and should convince others of the benefits of treatment.

**Chapter 8** looks back on the study and highlights the consequences. There is now proof that visual problems for people with intellectual disabilities are indeed an additional impairment in daily life and that treatment in this group is far from optimal. Improvement of treatment is difficult because of its complexity.

Decisions on interventions for people with intellectual disabilities are hampered because many people are involved (often also emotionally). They all must agree on the benefits, necessity and strategy of treatment. To improve the living environment it would help if government rules were made on light and use of color and contrast. Financial incentives are likely to support implementation.

Even after this study the existing protocols for screening and treatment of vision problems are still not sufficiently evidence based. However evidence for additional impairment by low vision is found and therefore it can be expected that early diagnosis followed by adequate treatment prevents problems.

For the daily practice in intellectual disability services and low vision centers this study had significant consequences. Once it became clear that treatment advice was poorly implemented, even during the study low vision centers started to find ways to improve implementation. By now a large implementation project has been started to find out how advising and treatment should be organized in order to reach the best results.

There are no reasons to suppose that treatment for people with intellectual disabilities can not take place the same way as for anybody else. Medical care for



low vision and blindness in this vulnerable group should be brought on the same level as that of the general population.



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**Samenvatting**



Dit proefschrift beschrijft de resultaten van een studie die is uitgevoerd naar aanleiding van eerdere onderzoeksresultaten van Jacques van Splunder.

Van Splunder onderzocht een representatieve steekproef van ruim 1600 mensen uit een populatie van ruim 9000 personen met een verstandelijke beperking. Volgens de criteria voor slechtziendheid en blindheid van de Wereldgezondheidsorganisatie (World Health Organization, WHO), bleek in deze groep ongeveer 15% van de mensen slechtziend te zijn en 5% blind. Van de ouderen met het syndroom van Down die ernstig verstandelijk beperkt waren, was zelfs vier van de vijf slechtziend of blind. Een andere belangrijke constatering van Van Splunder was dat het bij 40% van de slechtzienden of blinden die hij aantrof, helemaal niet bekend was dat zij niet goed konden zien. Zijn algemene conclusie was dat verstandelijk gehandicapten te weinig oogheelkundige zorg krijgen.

De Rotterdamse hoogleraar Gezondheidszorg voor mensen met een verstandelijke beperking, Heleen Evenhuis, besloot tot een vervolgonderzoek, waarin de vraag centraal staat wat het effect is, als bij volwassenen met een verstandelijke beperking slechtziendheid of blindheid wordt opgespoord en er vervolgens wél behandeling plaatsvindt. Dit proefschrift beschrijft de opzet, uitvoering en resultaten van dat onderzoek.

Als eerste moesten deelnemers aan het onderzoek gevonden worden: mensen met een verstandelijke beperking. Dat gebeurde via instellingen die zorg leveren aan deze doelgroep, zoals woonvoorzieningen, dagactiviteitencentra en sociale werkplaatsen. Het bleek echter niet eenvoudig om directies van deze instellingen te overtuigen van het nut van deelname. Van de vijftien benaderde instellingen wilden er slechts drie deelnemen en toezeggen het onderzoek te ondersteunen en de daaruit voortvloeiende behandeladviezen op te volgen.

Sommigen menen dat slechtziendheid voor verstandelijk beperkte mensen niet zoveel uitmaakt omdat ze toch al gehandicapt zijn. **Hoofdstuk 2** beschrijft het onderzoek naar de vraag of mensen die slechtziend of blind zijn, in het dagelijks leven anders functioneren dan mensen met ongeveer dezelfde ernst van verstandelijke handicap, die wel goed kunnen zien. Om dat te kunnen bepalen moesten meetinstrumenten worden gevonden. Voor mensen met een verstandelijke beperking bleken vragenlijsten te bestaan, en er waren andere vragenlijsten voor mensen die slecht kunnen zien. Vragenlijsten die geschikt zijn voor mensen met beide beperkingen bestonden echter niet. Uiteindelijk is besloten om twee bekende en betrouwbare vragenlijsten te gebruiken: de Sociale

Redzaamheidsschaal Zwakzinnigen (SRZ) van Kraijer (1994) en de Vragenlijst Over Gedrag (VOG) van Einfeld en Tonge (1995).

De SRZ is een veel gebruikte lijst, die meet hoe goed iemand in staat is alledaagse dingen zelf te doen: douchen, aankleden, opruimen, tafeldekken en dergelijke. De VOG is een lijst die oorspronkelijk is ontwikkeld om emotionele en gedragsstoornissen op te sporen bij kinderen met een verstandelijke beperking, maar deze blijkt inmiddels ook geschikt volwassenen. Beide lijsten vragen echter niet naar specifiek kijkgedrag zoals oog-hand coördinatie en visuele herkenning. Om ook daarover gegevens te verzamelen is voor dit onderzoek een nieuwe vragenlijst gemaakt in samenwerking met gedragswetenschappers van Sensis, Bartiméus, De Brink en Visio. Deze vragenlijst is – samen met de SRZ en de VOG – voor de 269 deelnemers aan het onderzoek ingevuld door hun groepsleiding. Pas daarna werd het gezichtsvermogen van elke deelnemer onderzocht. Van de 269 mensen bleken er 107 slechtziend of blind te zijn en dezen scoorden significant en klinisch relevant lager op zelfredzaamheid dan de niet-slechtzienden. Hun oog-hand coördinatie was slechter en hun mobiliteit was door de slechtziendheid geringer dan die van de niet-slechtzienden. Daarmee is aangetoond dat ook in deze groep slechtziendheid wel degelijk meer beperkingen veroorzaakt in het dagelijks leven.

**Hoofdstuk 3** geeft een overzicht van de literatuur over het behandelen van oogproblemen en slechtziendheid bij verstandelijk gehandicapten. Over dat onderwerp bestaan wel veel gevalbeschrijvingen, maar vergelijkende studies naar behandelresultaten op functioneel niveau bij deze doelgroep zijn er nauwelijks. Daar staat tegenover dat de meeste aandoeningen gewoon behandeld kunnen worden – eventueel met extra zorg direct voor en na een operatie. De conclusie uit het literatuuronderzoek is dan ook dat het voorschrijven en dragen van een bril, een staaroperatie, het afplakken van een lui oog of een operatie bij scheelzien ook uitvoerbaar is bij mensen met een verstandelijke beperking. Follow-up onderzoek is nodig, zodat op termijn kan worden aangetoond welk resultaat behandeling heeft gehad. Moderne technieken zoals het ‘ooglaseren’ zijn al op kleine schaal toegepast – vooral bij mensen die geen bril of contactlenzen verdroegen en vaak ernstig gehandicapt waren. Hierover worden goede resultaten gemeld.

Dat slechtziendheid bij mensen met een verstandelijke beperking vaak onbehandeld bleef was al bekend. Daarom was het in dit onderzoek extra van belang er voor te zorgen dat de aanbevolen behandeling ook daadwerkelijk werd uitgevoerd. **Hoofdstuk 4** beschrijft de manier waarop alle bij de aanbevolen

behandeling betrokkenen systematisch zijn geïnformeerd. Het behandeladvies was vaak complex. Het kon bestaan uit het voorschrijven van een bril, én meer licht op de werkplek, én een andere zitplaats in de woonkamer, én zonwering om verblinding tegen te gaan, én training van het team over de bejegening. Om zo'n samengesteld advies uit te kunnen voeren is het nodig dat alle betrokkenen goed samenwerken: groepsleiding, leidinggevendenden, gedragswetenschappers, artsen voor verstandelijk gehandicapten, technisch personeel en directie. Ook voor de staf van adviserende instellingen voor slechtzienden ligt hier een taak. In elke deelnemende instelling zijn al deze partijen uitgenodigd voor twee bijeenkomsten waarin gezamenlijk is geïnventariseerd op welke gebieden problemen waren te voorzien. Met de techniek van concept mapping (het in kaart brengen van ideeën) is een gedetailleerd overzicht gemaakt van alle geopperde praktische problemen. Vervolgens zijn deze besproken met de directies, om obstakels op tijd te signaleren en – waar mogelijk – op te heffen.

Na deze voorbereiding op het uitvoeren van het behandeladvies, werd van zestig personen in de behandelgroep geconstateerd dat zij slechtziend waren. Elk van hen kreeg een behandeladvies. **Hoofdstuk 5** beschrijft hoe de groepsleiding de gegeven adviezen beoordeelde en wat er vervolgens mee gebeurde. Over een periode van twaalf maanden na het advies vulden groepsleiders die de deelnemer goed kenden drie maal een vragenlijst in. Zij beantwoordden vragen als: Weet u dat er een advies is gegeven? Zo ja, weet u wat erin staat? Weet u waar u het advies kunt nalezen? Vindt u de adviezen zinvol? Voert u de adviezen uit? Zo nee, waarom niet? Uit de antwoorden bleek dat twee derde van de adviezen bekend was bij groepsleiders en dat ruim de helft van hen de adviezen wel zinvol vond. In slechts een derde van de gevallen verliep de uitvoering redelijk tot goed. Adviezen voor ernstig verstandelijk gehandicapten werden niet beter of slechter uitgevoerd dan die voor lichter verstandelijk gehandicapten.

**Hoofdstuk 6** geeft de resultaten van het onderzoek naar de centrale onderzoeksvraag: wat verbetert er na behandeling? In dit onderzoek werd een behandelgroep vergeleken met een controlegroep. Welke deelnemer in welke groep terecht kwam bepaalde het toeval, was dus willekeurig. Vandaar de naam gerandomiseerd gecontroleerd onderzoek. Dit type onderzoek wordt niet veel gedaan onder mensen met een verstandelijke beperking. Het geeft weliswaar de meest betrouwbare resultaten, maar is ook vaak zeer bewerkelijk en er kleven soms ethische bezwaren aan.

Bij iemand de diagnose van slechthoortheid stellen en vervolgens voor de controlegroep de behandeling daarvoor uitstellen in het belang van onderzoek, is ethisch moeilijk te rechtvaardigen. Dit probleem was in dit onderzoek te vermijden door gebruik te maken van een screeningsprogramma dat in alle deelnemende instellingen al was gestart. Medewerkers van het adviescentrum voor slechthoortheid kwamen regelmatig in de instellingen om mensen te screenen. Dat gebeurde bijvoorbeeld eens per maand gedurende een halve dag, waarbij dan steeds een beperkt aantal mensen werd onderzocht. Op die manier werd in de loop van een aantal jaren iedereen gescreend. Voor ons onderzoek werd afgesproken het screeningsproces van ruim een jaar in de beginperiode snel te doen, dan een periode niet te screenen en aan het eind van het jaar weer in korte tijd veel mensen te screenen. Zo werden in het onderzoeksjaar evenveel (of zelfs meer) mensen gescreend als andere jaren en kon toch een controlegroep gevormd worden van mensen bij wie aan het eind van het onderzoek het gezichtsvermogen bepaald zou worden, en aan wie dan – indien nodig – behandeling zou worden geadviseerd.

Over de mensen in de behandelgroep en die in de controlegroep werden op dezelfde tijdstippen vragenlijsten ingevuld. Uit Hoofdstuk 5 bleek al dat de uitvoering van de adviezen ernstig te wensen over liet: brillen werden vaak niet gedragen, staaroperaties lieten lang op zich wachten en de verlichting werd niet altijd verbeterd. Wij vonden geen verschil tussen de behandel- en de controlegroep. Daaruit kan echter niet worden geconcludeerd dat behandeling geen zin heeft. Als de behandeling vaak onvolledig is geweest, kan niet worden verwacht dat deze effect heeft.

**Hoofdstuk 7** geeft het commentaar op een Nederlands onderzoek: een retrospectief onderzoek naar het verloop van een verwijzing naar de oogarts – voor brilaanmeting of voor cataract – voor mensen met een verstandelijke beperking. Ook in dit onderzoek blijkt dat brillen lang niet altijd worden voorgeschreven en cataract operaties vaak niet worden uitgevoerd. Het commentaar gaat in op de vraag wie er eigenlijk verantwoordelijk is voor goede zorg bij slechthoortheid. Inmiddels zijn de verantwoordelijke directies zich wel meer bewust geworden van de grote aantallen mensen in hun instellingen die (nog) niet adequaat worden behandeld. Een complicerende factor is echter dat slechthoortheid een ‘onzichtbare’ aandoening is: de patiënt klaagt niet, lijkt niet ongelukkig en laat geen moeilijk gedrag zien. De omgeving ervaart geen problemen en dat houdt waarschijnlijk de motivatie voor behandeling laag. Ook familieleden zijn niet altijd gemotiveerd voor behandeling, zoals blijkt uit de uitspraak: “Mijn dochter



is geen type voor een bril". De conclusie luidt dat de arts voor verstandelijk gehandicapten of de huisarts in het opsporen van slechtziendheid een leidende rol zou moeten spelen, omdat deze de voors en tegens van behandeling kan beoordelen en zo nodig de betrokkenen kan overtuigen van het nut van behandelen.

**Hoofdstuk 8** kijkt terug op het onderzoek en gaat onder andere in op de gevolgen die het heeft gehad. Aangetoond is dat slechtziendheid voor mensen met een verstandelijke beperking inderdaad een extra beperking betekent en dat er in deze groep nog veel te weinig wordt behandeld. Het doorvoeren van verbeteringen blijkt lastig vanwege de complexiteit van de behandelingen.

Ook komen beslissingen over mensen met een verstandelijke beperking moeilijk tot stand, doordat er heel veel partijen bij zijn betrokken. Die moeten het allemaal eens zijn over nut, noodzaak en aanpak van een behandeling. Voor het verbeteren van de directe leefomgeving zou het kunnen helpen als de overheid aan instellingen regels stelt op het gebied van verlichting en kleurgebruik. Ook van financiële prikkels is elders reeds aangetoond dat ze de implementatie bevorderen.

De bestaande richtlijnen voor het opsporen en behandelen van slechtziendheid zijn ook na dit onderzoek nog niet voldoende wetenschappelijk onderbouwd. Nu er meer bewijs is dat slechtziendheid een extra handicap is, kan worden verwacht dat een vroegtijdige diagnose die gevolgd wordt door behandeling, veel problemen kan voorkomen.

Voor de dagelijkse praktijk in de instellingen heeft het hier beschreven onderzoek duidelijke gevolgen gehad. Nadat bleek dat de adviezen slecht werden uitgevoerd, gingen de adviserende instellingen nog tijdens het onderzoek aan de slag om verbeteringen door te voeren. Inmiddels is er zelfs een groot implementatie-onderzoek gestart om te achterhalen hoe advisering en behandeling zo georganiseerd kunnen worden dat er wel gebeurt wat nodig is.

Voor de veronderstelling dat behandeling van slechtziendheid bij mensen met een verstandelijke beperking niet kan plaatsvinden zoals voor ieder ander, bestaan geen aanwijzingen. De medische zorg voor slechtziendheid en blindheid in deze kwetsbare doelgroep zou daarom zo snel mogelijk op hetzelfde niveau gebracht moeten worden als dat van de algemene bevolking.



## Manuscripts based on the studies presented in this thesis

Sjoukes L, Koot JM, Kooijman AC & Evenhuis HM. Does visual impairment lead to additional disability in adults with intellectual disabilities? A cross-sectional study. *Journal of Intellectual Disability Research* (in press).

Sjoukes L, Harteloh PPM & Evenhuis HM. Brief research report. Is concept-mapping an effective method of introducing complex interventions into intellectual disability services? *Journal of Policy and Practice in Intellectual Disabilities*. 2006 June; 3 (2) 133–135.

Sjoukes L, Kooijman AC, Koot JM & Evenhuis HM. Rehabilitation of low vision in adults with intellectual disabilities: The influence of staff. *Journal of American Research of Mental Retardation* (in press).

Sjoukes L, Kooijman AC, Koot JM & Evenhuis HM. A randomized controlled trial of interventions for visual impairment in adults with intellectual disabilities (submitted).

Evenhuis HM & Sjoukes L. Inadequacies in the treatment of visual impairment in people with an intellectual disability: Who is responsible? (Comment) *Nederlands Tijdschrift voor Geneeskunde*. mei 2008 152; (18) 1034–35.



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## Curriculum vitae

Liesbeth Sjoukes is geboren in 1950 in Bandoeng, Indonesië. Ze volgde de zesjarige meisjes-HBS in Utrecht en behaalde in 1968 het eindexamen HBS-B. Aansluitend studeerde ze van 1968-1975 geneeskunde in Utrecht.

Als arts werkte zij tot 1979 in het Dr. J. N. Voorhoeve verpleeghuis te Utrecht. In de periode 1976-1979 was ze als docent verbonden aan de opleiding tot ziekenverzorgende in Utrecht en was ze een van de presentatoren van het KRO-televisieprogramma "Daar vraag je me wat".

Van 1994 tot 2001 werkte ze als arts voor verstandelijk gehandicapten (AVG) bij Zonnehuizen in Zeist. Ze volgde de voorloper van de huidige AVG opleiding bij Henny Schroijenstein Landman-de Valk.

Van 2001-2005 werkte ze aan het onderzoek bij Heleen Evenhuis.

Eind 2005 keerde ze weer terug naar Zonnehuizen, nu om te werken op de afdeling Aventurijn, die psychiatrische dagbehandeling biedt aan kinderen met een lichte verstandelijke beperking.

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