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Visual impairments in people with severe and profound multiple disabilities: an inventory of visual functioning

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Abstract

Background The prevalence of visual impairments in people with severe and profound multiple disabilities (SPMD) is the subject of considerable debate and is difficult to assess.
Methods In a typical Dutch care organization, all clients with SPMD ($n = 76$) participated in the study and specific instruments adapted to these clients (requiring a minimum of cooperation) were used to measure visual acuity, the visual field, binocular vision, contrast sensitivity, refractive errors and visual functioning behaviour.
Results We found an unexpected 92% of clients with SPMD to have visual impairments. Previously, only 30% were known to have visual problems. None of the persons observed had normal visual acuity. Subnormal visual acuity was the best result. The severity of the visual impairment was related to the severity of the intellectual disability. In addition to the problem of acuity, impairments in the visual field, impaired contrast sensibility and impaired binocular functioning were found, as well as impaired visual attention, fixation and following. In 22% of the clients observed, refractive errors were found and glasses were advised.
Conclusions Consequences for caregiving and for modifications of the environment were discussed.

Keywords intellectual disability, multiple disabilities, prevalence of visual impairment, visual functioning

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Introduction

A number of studies [in the Netherlands led by Evenhuis (1995) and Evenhuis *et al.* (2001)] have shown that the prevalence of visual impairments in people with an intellectual disability (ID) must be higher than the 2% in the rest of the population. This higher prevalence is assumed, although a diagnosis of visual impairment has not yet been made for most people with ID. This under-diagnosis may be explained by failure to complain about visual problems, while family or professionals involved may interpret atypical behavioural patterns as a result of the intellectual impairment overshadowing the visual impairment. Furthermore, it is difficult to examine the vision of a person with an ID.

Visual impairment is defined in terms of two visual functions: visual acuity and visual field. Visual acuity is the ability to distinguish details. The World Health Organization (WHO) norm for average visual acuity is 1.0. The visual field is the area in which objects can be seen in the peripheral field while the eye is focused on a central point. The average visual field is 180°. According to WHO criteria, a visual acuity of less than 0.3 and more than 0.05, or a visual field of between 30° and 10° indicates visual impairment. According to the same criteria, a visual acuity of less than 0.05 or a visual field of less than 10° constitutes blindness. Impairments of visual functions can be caused by ocular characteristics (e.g. refractive errors, injuries to the retina, cataracts and high ocular pressure) as well as by cerebral characteristics. Damage to the cerebral visual system, visual cortices and the visual pathways (optic nerve) can diminish visual acuity and restrict the visual field.

In recent years, several studies on visual impairments in intellectually disabled people have been published. Many of these are collected in a review by Warburg (2001). Alarming figures concerning prevalence of blindness and serious visual impairment in this group are given, ranging from 5% to 78%. Reasons for the discrepancies may be the small groups of subjects, different selection procedures and different assessment methods used. However, there is an obvious trend: prevalence is related to age and to the severity of the ID. The Dutch prevalence study carried out by van Splunder (2003) is fairly extensive. He found visual impairment in 2.2% and blindness in 0.7% in people less than 50 years of age with a mild degree of ID and no Down's syndrome. In people older than 50 years, with a profound ID and Down's syndrome, he found visual impairment in up to 67% and blindness in up to 16.7%. He extrapolated his results to the total population of people with an ID in the Netherlands: 14% had visual impairment and 5% had blindness. In people with Down's syndrome age seems to correlate more strongly with visual impairment; whereas in people without Down's syndrome the degree of ID has a stronger correlation with visual impairment. Van Splunder also found that in more than 40% of the visually impaired (43% with visual impairment and 38% with blindness), the diagnosis of either visual impairment or blindness had not previously been made. Van Splunder's firm conclusion was that persons with a severe or profound ID should be considered visually impaired until evidence to the contrary is found.

The higher prevalence of visual impairment in people with an ID is partly triggered by the ophthalmologic problems related to specific syndromes (Down's syndrome, Angelman syndrome, tuberous sclerosis, fragile X syndrome, Prader–Willi syndrome). Curtailment in the lower half of the visual field has previously been described by Jacobson *et al.* (1996) in children with periventricular leucomalacia, polycystic areas in the white matter of the brain around the ventricles. In several syndromes, Marfan syndrome, Rubella, Cri du chat, the symptom of strabismus (crossed eyes) is mentioned (Hou *et al.* 1999). For the rest, visual impairments in this group may be ascribed to cerebral genetic impairments or brain injuries. Vlaskamp (2003) regards persons with a serious or profound ID as persons with multiple disabilities, because they frequently suffer from co-morbid functional, motor and sensory disorders, and disorders such as epilepsy, gastro-oesophageal reflux and challenging behaviour. De Jong (1986) speaks of multiple disabilities if at least two essential perceptual, behavioural or communicative functions are absent that cannot be compensated for by other functions.

In the Netherlands, many organizations nowadays monitor their clients' visual functioning. That is why the organization that participated in this study, which provides care and housing to almost 2000 clients in various locations, asked the first author to monitor the visual functioning of its clients, starting with the seriously and profoundly multiply disabled. This study gives an inventory of the visual functioning of this particular group, looking at visual acuity, visual field, contrast sensitivity, binocular vision, visual behaviour, refraction errors and the frequency with which glasses were prescribed. In this study, we are also interested in the relationship between visual functioning and age, cerebral palsy, epilepsy, and the use of psychiatric medication and anti-epileptic drugs.

Method

Subjects

All clients (43 male; 33 female) with severe and profound intellectual and motor disabilities living in a typical Dutch care facility participated. Sixty-five per cent of these clients use specially made wheelchairs; the other 35% are able to walk a short distance. Twenty per cent are unable to make any focused movements. None are able to communicate other than non-verbally. People with acquired brain damage and people with dementia were excluded. Informed consent was acquired from the families of all clients.

Instruments and procedure

Because of the seriousness of ID, it was only possible to use test materials that required minimum cooperation. Some visual functions could not therefore be examined. For example, colour vision can only be tested if a person has the ability to match. Visual screening was performed on-site by an orthoptist. The time set aside for screening was extensive and it was performed in a playful manner. All clients were accompanied by a significant person from their social network, who was asked if and in what way the client's behaviour was different during screening. If the answer was positive, a new appointment was made for another screening day. If visual problems were found, an additional live observation was performed to add to the reliability of the observations made on the screening day.

Visual acuity

Resolution (grating) acuity testing was measured by two preferential looking tests: the Teller acuity cards (Teller 1972) and the Cardiff cards (Woodhouse *et al.* 1992). These tests assume that an individual who is able to discern an image on one side of a card will look towards that side. Subjects are presented with Teller acuity cards showing a black-and-white striped area ('gratings') on one side, and a uniform grey on the other side. The gratings are of different widths and are positioned on the left, and sometimes on the right side of the card. If the subject is able to see and discern the pattern, he or she will prefer to look at the striped area. The critical width at which the subject can no longer see the stripes, related to the distance at which the card was presented, is used as the measure for grating acuity, which was expressed in cycles/degrees. The Cardiff test cards present popular figures in the same way.

Visual field

Testing the visual field was performed by using Stycar graded balls and the confrontational method (Sheridan 1973): the subject's attention is attracted while a ball secured to a rod is made to enter the visual field by someone standing behind the subject. The ball is moved from various different positions from the periphery towards the centre of the visual field, and an observer monitors the moment when the subject first catches sight of the ball. This test cannot find blind spots in the centre of the visual field. This study speaks of a diminishing visual field when it is substantial and has been demonstrated in different situations.

Binocular vision

Both eyes are positioned correctly and work together (stereovision). Impairment is assessed if only one eye is used or both eyes are used alternately.

Contrast sensitivity

How faded can an image be before it becomes indistinguishable from a uniform field? This variable was tested with the low contrast Hiding Heidi faces (Hayvärinen 1998), by using the preferential looking technique. Faces printed in lines varying from 100% to 1.2% contrast are presented to the subject. By observing the direction of gaze, it is possible to see whether the visual information is perceived. A diminishing of contrast sensitivity is established if the subject does not perceive a contrast difference of 5%, which means that he or she will have difficulty in perceiving facial expressions.

Refractive errors

An autorefraction meter (Nikon Retinomax, Nikon Inc., Melville, NY, USA) measures refractive error. As the subject fixates an image on a screen, the meter automatically calculates if glasses are needed. This is an indication for further ophthalmologic investigation. Cycloplegic eye drops cause a loss of accommodation, making an exact measurement of refraction possible. Measurement of refraction, investigation of media (e.g. cataract) and a funduscopic examination were performed by slit lamp biomicroscopy to detect problems in the retina and the optical nerve.

Only if a visual impairment was diagnosed ($n = 61$) did an observation of behaviour follow to assess visual functioning behaviour, attention-span during visual activities, ability to fixate different moving targets, and the quality of eye–hand coordination.

Results

All subjects participated (no non-response, after informed consent had been obtained from the families). The majority of the clients had a profound ID (83%), cerebral palsy (61%) or epilepsy (71%). Average age was 33 years (SD = 12.6; age groups 4–10 years: $n = 3$; 11–20 years: $n = 11$; 21–30 years: $n = 24$; 31–40 years: $n = 29$; 41–50 years: $n = 3$; 51–74 years: $n = 6$). Because some measurements were unreliable, certain data are based on different numbers of subjects.

The prevalence of impaired visual acuity was very high at 92% (Table 1). In none of the clients was normal visual acuity found; subnormal visual acuity was the best result.

A very high prevalence of severe and profound visual impairment and blindness was found. Most of the clients could see and follow a light in a dark area. Only two people did not react to a strong light. Apart from impaired visual acuity, a curtailment of the visual field was found in more than half of the clients (51%). Curtailments were often found at the left or right side and in the lower half of the visual field [curtailments in this area have previously been described by Jacobson *et al.* (1996) in children with periventricular leucomalacia: polycystic areas in the white matter of the brain around the ventricles]. Eighty per cent of the clients were found to have impaired contrast sensitivity and 84% were not able to use both eyes (binocular vision). In most of these clients, suppressed amblyopia (lazy eye), caused by crossed eyes, was found. Sixty-one per cent of the clients diagnosed as seriously visually impaired showed problematic visual behaviour consisting of a problematic attention span during visual activity, and difficulty in fixating and following objects. The visual impairments were partly caused by ophthalmologic problems. Refractive errors partly explain the diminished visual acuity. By using the autorefraction meter without cycloplegic eye drops, refractive errors were found in 60% of the clients. In 22%, these errors led to a further examination using cycloplegic eye drops if it was thought that the use of glasses might be helpful. After this examination, glasses were prescribed in all cases.

The severity of the visual impairment was significantly worse in clients with profound ID than in clients with a severe ID: visual acuity (χ^2 24.2, d.f. = 3, $P < 0.01$), contrast sensitivity (χ^2 10.8, d.f. = 1, $P < 0.01$) and binocular vision (χ^2 7.7, d.f. = 1, $P < 0.01$). Curtailment of the visual field as well as impairments of visual attention, fixation and following of targets showed no significant relation to intellectual functioning. Although there was no significant correlation between severity of the ID and refractive errors, significantly more people with severe ID were prescribed glasses compared with people with profound ID (χ^2 4.1, d.f. = 1, $P < 0.04$).

Clients with cerebral palsy (61%) did not experience greater visual problems with regard to visual acuity, visual field, contrast sensitivity and binocular vision. They showed significantly more serious impairments in visual attention, fixation and following (χ^2 9.8, d.f. = 1, $P < 0.01$).

Clients with epilepsy (71%) experienced significantly more often severe problems in visual acuity (χ^2 10.4, d.f. = 3, $P < 0.02$), and in visual attention, fixation and following (χ^2 4.5, d.f. = 1, $P < 0.03$).

Seventy-two per cent of clients were taking psychiatric medication or anti-epileptic drugs. A significant correlation was found between severity of visual

Table 1 Percentage of people with impaired visual acuity ($n = 74$; for two clients measurements were not reliable)

	Visual impairment (VI)		Blind (WHO)	
Subnormal < 1.0	Moderate VI < 0.3– > 0.1	Severe VI < 0.1	Profound VI < 0.05	Blind
8%	27%	40%	22%	3%

WHO, World Health Organization.

acuity impairment and curtailment of the visual field on the one hand and the use of both types of medication on the other (χ^2 11.9, d.f. = 3, $P < 0.01$ and χ^2 4.0, d.f. = 1, $P < 0.04$, respectively).

Previous studies have found that age is positively related to visual impairments. In this study, we found significantly more serious problems, especially in relatively younger people: visual acuity (ANOVA, $F = 4.2$, d.f. = 3, $P < 0.01$; least serious problems mean age 45 years and SD = 18, most serious problems mean age 26 years and SD = 9) and binocular vision (ANOVA, $F = 5.1$, d.f. = 1, $P < 0.03$; no problems mean age = 38 years and SD = 12, serious problems mean age = 29 years and SD = 13). Further analysis showed that (mean age = 29 years, SD = 11), the prevalence of profound ID was higher in younger people than in older people (mean age = 38 years, SD = 16) (ANOVA, $F = 6.4$, d.f. = 1, $P < 0.01$). This may explain the different results.

Discussion

Although all clients were recruited from the same care organization, we have not found any reason why the selected group would be any different from clients cared for by other Dutch organizations. The lack of non-response reinforces this conclusion. Despite the difficulties in monitoring the vision of this group, the many serious problems in visual functions found are unmistakable and should be dealt with in care provision. Before this study, a visual problem was diagnosed in only 30% of cases according to clients' personal files. We found a prevalence of 92% and, to paraphrase van Splunder (2003), people with serious and profound multiple disabilities should all be perceived as people with serious visual problems until this is proven otherwise. A correlation was shown between visual functioning and the severity of the ID.

Our results may be used to optimize or redirect care for this group. When advising parents and caregivers about sensitive communication and adaptations in the environment, it may not be enough to make a distinction between the visually impaired and the blind. The world of people with an acuity of 0.25 is different from those with 0.1. Nor is the concept of blindness particularly precise, because the WHO criterion of 0.05 acuity allows people to look at and enjoy strong visual stimuli, that is, those stimuli that are used during 'snoezelen'. A diagnosis of blindness may result in withdrawal of activities like 'snoezelen'. In addition, it will make a great difference if the impairment is based on acuity or on the size of the visual field. Those who provide care to people with serious or profound multiple disabilities need to be trained, **and** the high prevalence of persons with problems in contrast sensitivity make personalized environments and, in particular, communication necessary, because contrast sensitivity is strongly related to perception of facial expressions. Proximity, adapted illumination and make-up accentuating facial expressions may be important. The many specific problems in the visual field should encourage caregivers to evaluate their presentation of materials. If the lower half of the visual field is affected, materials presented on a table may not be seen. Caregivers may not notice the problems of the moderately visually impaired (acuity 0.3–0.1), because an acuity of between 0.3 and 0.1 allows for good perception of a drinking cup and many toys. Even small pieces of chocolate popcorn can be seen from a short distance with an acuity of 0.1. Many people only explore their close environment, and caregivers must be sensitive in seeking proximity. Only when signs or photographs are used and perception of details is required will the visual impairment become evident. It will be clear that the different impairments in acuity, visual field and contrast sensitivity in combination with the impaired visual behaviour should lead to individual advice.

Further research is necessary so that protocols can be developed for people with profound ID who are getting used to wearing glasses. Effects can then be monitored and changes in behaviour analysed. The correlation found between epilepsy and medication on the one hand and visual acuity on the other is interesting and needs to be explored further.

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