

## Differentiating characteristics of deafblindness and autism in people with congenital deafblindness and profound intellectual disability

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### Abstract

**Background** In persons with deafblindness, it is hard to distinguish autism spectrum disorders from several deafblind specific behaviours caused by the dual sensory impairments, especially when these persons are also intellectually disabled. As a result, there is an over-diagnosis of autism in persons who are deafblind leading to unsuitable interventions.

**Methods** Autism as specified by the DSM-IV was studied in 10 persons with congenital deafblindness with profound intellectual disabilities. Behaviours of people with deafblindness and autism ( $n = 5$ ) and of people with deafblindness without autism ( $n = 5$ ) were observed in a semi-standardised assessment.

**Results** All people with deafblindness showed impairments in social interaction, communication and language. In contrast to persons without autism, people with deafblindness and autism showed significantly more impairments in reciprocity of social interaction, quality of initiatives to contact and the use of adequate communicative signals and functions. No differences between the

groups were found for quantity and persistence of stereotyped behaviour, quality of play and exploration and adequate problem-solving strategies.

**Conclusions** This study indicates that there are some possibilities to differentiate autism from behaviours specific for deafblindness. It also confirms the large overlap in overt behaviours between people with deafblindness and persons with autism.

**Keywords** autism, concomitant behaviour, deafblindness, differential diagnosis, intellectual disabilities

### Introduction

Professionals in clinical practice are often consulted about autism in people with deafblindness. This is not surprising as the prevalence of autism seems to be positively correlated to hearing impairments (Carvill 2001), visual impairments (Cass 1998) and intellectual disabilities (ID) (De Bildt *et al.* 2005). Subsequently, it is to be expected that people with congenital deafblindness have a higher risk of being autistic, too. However, several authors have stated that there is an over-diagnosis of autism in persons with sensory impairments, because of topographical similarities in behaviours but differences in the

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underlying mechanisms or processes that cause these behaviours (Hobson *et al.* 1997; Cass 1998; Knoors & Vervloed 2003; Andrews & Wyver 2005). In the current study, the characteristics of people with deafblindness and ID with or without autism are compared with study similarities and differences in overt behaviours in order to correctly identify people who are deafblind and intellectually disabled with or without autism.

In deafblindness, there is a combination of two or more sensory disabilities with an early onset for which education or intervention developed and suitable for children with one disability is not applicable. In people with congenital deafblindness, it is primarily the development of communication and language that is affected (Carvill 2001; Gense & Gense 2005), although other developmental areas are also likely to be affected such as social-affective development (Gense & Gense 2005), cognitive development (Van Dijk *et al.* 1993) and motor development (Van Dijk & Janssen 1993; Gense & Gense 2005). People with deafblindness often show atypical or stereotyped behaviours (Murdoch 2000; Hartshorne *et al.* 2005), many of which are also shown by persons with severe ID or with psychiatric disorders such as autism (Van Dijk *et al.* 1993).

#### Characteristics of autism and deafblindness

With regard to autism, the DSM-IV-TR (American Psychiatric Association 2000) describes three main domains of problematic development: (1) qualitative impairments in social interaction; (2) qualitative impairments in communication; and (3) restricted patterns of behaviour, interests and activities. With regard to qualitative impairments in social interaction in autism, there might be problems in initiating social interaction and contact. In general, there is a lack of empathy and social or emotional reciprocity (Frith 2003; Gense & Gense 2005). Persons with autism show a complete absence or lack of spontaneous attempts to share attention and pleasure and to involve others in their activities (Frith 2003; Gense & Gense 2005). They also show deficiencies in understanding social meanings of interactions and are not able to participate in social situations adequately (Frith 2003). These behaviours show some similarities with social behaviours in people

who are deafblind and who often show withdrawal from and a general lack of social interaction (Van Dijk 1995). Initiatives for contact are mostly concerned with the satisfaction of personal needs or attempts to disengage social contacts (Van Dijk & Janssen 1993). Mutual looking or joint visual attention is often lacking, although they might be able to share attention and able to involve others in their activities by compensatory methods (Van Dijk & Janssen 1993; Dale 2005; Gense & Gense 2005; Jordan 2005). Many of these behaviours are the result of the dependency on others for taking the initiatives for social contact (Knoors & Vervloed 2003). People with deafblindness experience common situations often as fearful, because they lack the proper information to interpret the situation correctly. To avoid these fearful situations, they might withdraw from social contact completely, which in turn leads to social deprivation (Van Dijk 1995).

The second characteristic of autism, qualitative impairments in communication, can also be found in people with deafblindness. Persons with autism show impairments in the use of nonverbal behaviours such as eye-to-eye gaze, social smile, body posture and facial expressions. Besides, they also show difficulties in perceiving and understanding nonverbal communicative signals such as facial expressions, gestures and intonation (Frith 2003). For people with deafblindness, it is extremely difficult to perceive good models of nonverbal behaviour. Social smiles and facial expressions are easily missed (Van Dijk & Janssen 1993), and eye-to-eye gaze might be seriously restricted by the visual impairment (Jordan 2005). Some persons with autism perceive communication fragmentally (Noens 2004), as do some persons who are deafblind. However, the latter do so because visual or auditory information is missed, not because communicative signals are not perceived or understood. Many persons with autism show a delay in or total lack of development of spoken language (Frith 2003; Gense & Gense 2005). In persons with autism who show reasonable syntactic and semantic skills, pragmatic skills are often impaired (Frith 2003). In persons with ID, communication is often at a prelingual level, and only primitive signals are used to express oneself (Noens 2004). Usually, the social use of language is also impaired (Kraijer

2003). For people with deafblindness, spoken language is not readily accessible because of the hearing impairment; consequently, their spoken language is delayed or even absent. Moreover, in people with deafblindness the opportunities to communicate are restricted to persons who are trained in (tactile) sign language. Communication of many persons with deafblindness often stays at the prelingual level for a long time, and many of them will never reach a symbolic level of communication. This is especially the case when the person is also intellectually disabled (Van Dijk & Janssen 1993).

The third clinical criterion of autism refers to restricted patterns of behaviour, interests and activities. Persons with autism often show stereotyped and repetitive movements with their body (Frith 2003; Hartshorne *et al.* 2005) and objects (Gense & Gense 2005). The nature of these movements is very persistent and complex. Attempts to stop these behaviours often result in extreme resistance to change, and this behaviour will increase in intensity when another person takes initiatives for interaction (Gense & Gense 2005). Persons with autism often show rigid adherence to behavioural routines and rituals (Frith 2003; Gense & Gense 2005). Also, many persons with blindness and deafblindness show stereotyped and repetitive behaviours with their bodies and objects (Murdoch 2000; Hartshorne *et al.* 2005). With age and with increased opportunities to communicate, to initiate and to maintain interactions and explorations, these behaviours will usually decrease (Van Dijk & Janssen 1993; Murdoch 2000).

### Differential diagnosis

Because a neurochemical or neurophysiological base of autism has not been identified yet and brain maladaptations found in people with autism are still inconsistent (Noens 2004), the diagnosis of autism is based on behavioural characteristics. If behaviours are topographically the same, it is useful to know whether the underlying factors that cause the behaviours are also the same. In other words, one wants to know whether there is equifinality, diverse paths or factors associated with the same outcome. With regard to the present study, this question could be: is the behaviour caused by the presence of

autism or is it a result of the sensory impairments? Some data from the literature illustrate this problem.

Smith *et al.* (2005) mentioned the risk that atypical behaviour in people with deafblindness is unjustly attributed to the multi-sensory impairments, consequently masking a diagnosis of autism. On the other hand, these same atypical behaviours can easily be confused with psychopathological disorders such as autism (Freeman & Groenveld 2000). Van Dijk (1982) raised the idea that particular organic conditions and insecure attachments may lead to autistic-like behaviours in persons with congenital rubella syndrome. According to Van Dijk & Nelson (1996) and Rødbroe (2001), the stereotyped and repetitive behaviours of people with deafblindness can be seen as consequences of sensory and social deprivation and lack of compensatory strategies. Note that in persons with autism these same behaviours are often attributed to a coping mechanism for overstimulation (Gense & Gense 2005) or the result of impairments in executive functioning (Frith 2003). Concomitant ID further complicate the picture. Many people with deafblindness have brain damage, whether congenital or not, which directly affects their behaviour and motor development and might cause problems in perception.

One can readily understand that, when it is hard to differentiate between characteristics of autism and deafblindness in controlled studies, it will be even harder to do so in clinical practice, especially in individuals with additional ID. Existing standardised tests, questionnaires and developmental scales are neither reliable nor valid for use with people with deafblindness (Westland *et al.* unpublished data). The main reason is that these instruments do not take into account the combined effect of multiple disabilities (Nelson *et al.* 2002; Smith *et al.* 2005).

Based on the literature, the conclusion is justified that differentiating autism in people with deafblindness is difficult or even impossible. However, it is also scarcely tried before. The current study is an attempt to objectify autism-specific behaviours in people with deafblindness. Aim of the study is to explore the possibilities for diagnosing autism correctly in people with deafblindness. Because deafblindness is often accompanied by ID and the

questions and needs in assessment are the highest within this group, this study has been conducted with deafblind persons with ID as well. The research question is: 'What are the specific behaviours by which people with deafblindness who are autistic or not can be distinguished from each other?'

## Methods

### Participants

Congenitally, deafblind participants with ID were recruited from a large institute for people with deafblindness. The research sample had to be representative for persons with deafblindness and autism and should include a control sample of persons with deafblindness without autism in order to find differences and similarities in behaviours of persons with deafblindness with or without autism. As unambiguous diagnostic criteria for autism in people with deafblindness are lacking, consensus in judgments about the presence or absence of autism was used. A panel consisting of a psychiatrist, psychologist and an independent expert in deafblindness all rated the participants independently. These experts had all ample experience with deafblind people – see Brown *et al.* (1997) for a rationale for using an expert judgment on autism instead of assessment instruments. A total sample of 95 congenitally deafblind persons were judged by the panel. For 63 (66%) persons, consensus about the presence of autism was reached ( $n = 20$  autism present and  $n = 43$  autism absent); for 32 (34%) persons, no consensus could be reached.

In order to participate, the following additional criteria had to be met:

- Binocular visual acuity  $<0.3$ , determined by an ophthalmologist or orthoptist;
- Bilateral severe or profound hearing loss: Fletcher index  $>60$  dB and/or auditory neuropathy, determined by an audiologist; and
- Cognitive functioning less than a developmental age level of 24 months, determined by an educational or developmental psychologist.

Based on examination of medical and psychological records, 25 out of 63 persons with congenital deafblindness and consensus on the presence or absence of autism satisfied the above mentioned criteria.

Parents or caregivers of 17 persons did consent to participation. After further analysis of records and after considering the severity of motor disabilities, six persons were excluded from the study. The assessment of one participant had to be stopped because of personal circumstances. Leaving 10 persons with deafblindness who participated after informed and written consent, all of them had severe ID (see Table 1). Of those 10 persons, five had been diagnosed with autism, and five did not show any sign of autism.

The mean age of the participants with autism was 18 years (ranged 7–28), and the mean age of the deafblind participants without autism was 15 years (ranged 5–23). Most of the participants with autism were male (80%), and most of the deafblind participants without autism were female (80%). One of the participants with autism did not have any functional residual vision. Other participants with autism and all participants without autism had some residual vision. Hearing loss was 45 dB in one client with auditory neuropathy. All other participants had hearing losses of at least 60 dB (ranged 60–110 dB; see Table 1 for an overview).

### Instrument

The instrument – 'observation of characteristics of autism in persons with deafblindness' (O-ADB) – was developed for this study. The O-ADB contains concepts that were found in the literature on autism-specific behaviours. The O-ADB was developed with the help of both experts in deafblindness as well as in autism. The items and norms of the O-ADB were based on the Autism Diagnostic Observation Schedule of Lord *et al.* (2006), the Autism Screening Instrument for Educational Planning of Krug *et al.* (1980), the Autism Diagnostic Interview-Revised of Le Couteur *et al.* (2003) and the Van Dijk Approach to Assessment (also known as 'Hands-on' Assessment; Nelson *et al.* 2002). This resulted in a semi-standardised observation-instrument. Standardisation consisted of: (1) well-described target behaviours and activities; and (2) the use of a standard set of materials suited for persons with sensory impairments and of a hierarchy in prompts by the examiner (Lord *et al.* 2006). To increase the probability that the persons with deafblindness would show their competencies and

**Table 1** Characteristics of deafblind participants

Participant	Diagnosis autism*	Age (years)	Sex	Hearing loss†	Visual acuity‡ / visual field loss	Aetiology/syndrome
1	+	17	Male	65 dB	0.15/-	Goldenhar
2	+	23	Male	>95 dB	Light perception/-	Congenital rubella
3	+	15	Male	75-85 dB	0.1/-	CHARGE
4	+	7	Male	45 dB‡	0.15/+	CHARGE
5	+	28	Female	110 dB	0.25/+	Congenital rubella
6	-	23	Female	108 dB	0.2/-	Congenital rubella
7	-	5	Male	65-80 dB	0.1/?	Zellweger
8	-	22	Female	70 dB	0.01-0.03/-	Prematurity
9	-	14	Female	60-75 dB	0.1-0.3/-	West
10	-	12	Female	80 dB	0.1/-	Trisomie 22 mosaic

\* Diagnosis autism by consensus judgment (+) or no doubt of absence of autism by consensus judgment (-).

† Hearing loss expressed by Fletcher index of best ear, i.e. mean hearing loss at 500, 1000 and 2000 Hz.

‡ Vision loss expressed by binocular visual acuity/impairments of visual fields; +, present; -, absent; ?, unknown.

§ In spite of the moderate hearing loss, this participant was included in the experimental group, because observations of auditory functioning showed that this participant did not use his residual hearing at all and was therefore suspected to have auditory neuropathy.

skills, different ways for initiating interaction were used. Initiatives and interests of the person with deafblindness were followed with a tactile approach (Nelson *et al.* 2002).

Scoring categories for the O-ADB are described in Table 2 and concerned quality and frequency of target behaviours. Higher scores depict higher quality of the behaviours and lower scores, more autism-specific behaviours. Behaviours were scored that were directed either to the unfamiliar examiner or to a familiar person present during the assessment. The internal consistency for 27 items of the O-ADB was high (Cronbach's  $\alpha = 0.95$ ). Intra-class correlations were used to measure the inter-rater agreement. After correction for miss values by deleting the items 'eye gazing' and 'breaking stereotyped behaviour', there was sufficient and significant inter-rater agreement, except for stereotyped behaviour (see Table 2). A more-extensive description of the O-ADB can be found in Hoevenaars & Antonissen (2007) and in Hoevenaars *et al.* (2008).

### Procedure

The assessment took place in an observation room in the participants' institute. All observations lasted between 35 and 60 min. The first two authors, both

educational psychologists, carried out the assessments. Each of them assessed five participants, both with and without autism. A parent, teacher or caregiver was always present during the observation and, if necessary, was asked to support the educational psychologist, e.g. to communicate with the participant. All assessments were recorded in video. Each tape was subsequently observed and scored by four observers with the help of a standardised scoring protocol. The first two authors scored all video tapes. Each video tape was further observed by one of four experts in deafblindness and one of three experts in autism. The experts received written instructions on the scoring and were not informed whether the client was autistic or not. For each participant, averages were calculated per item based on the scores of the four different observers. Subsequently, these average scores were accumulated into a total behaviour score. Because of many missing values for the items 'breaking stereotyped behaviour' and 'eye gazing', these items were left out in the calculation of the total scores. The missing values on the item 'breaking stereotyped behaviour' were the result of the conditional character of this item. This item was only administered whenever a participant showed continued stereotyped behaviour (with his own body or with objects) for at least 2 min. The missing values for

**Table 2** Target behaviours, categories and their reliability of the observation of characteristics of autism in persons with deafblindness

<p><b>Openness for contact (0.70/0.49)*:</b></p> <ul style="list-style-type: none"> <li>• Openness for initiatives in contact;</li> <li>• Pleasure in contact; and</li> <li>• Acceptance of bodily contact.</li> </ul>	<p><b>Communicative signals/functions (0.92/0.89):</b></p> <ul style="list-style-type: none"> <li>• Initiative to contact;</li> <li>• Initiative to bodily contact; and</li> <li>• Communicative function: functional or social instrumental way of contact.</li> </ul>	<p><b>Stereotyped behaviour (0.63/0.23):</b></p> <ul style="list-style-type: none"> <li>• Stereotyped behaviour with own body;</li> <li>• Stereotyped behaviour with objects; and</li> <li>• Response on breaking stereotyped behaviour<sup>†</sup>.</li> </ul>
<p><b>Reciprocity/joint attention (0.91/0.77):</b></p> <ul style="list-style-type: none"> <li>• Following joint attention for some object or person by eye gazing, pointing and acting;</li> <li>• Initiative to joint attention;</li> <li>• Turn taking;</li> <li>• Anticipation of acts in routines;</li> <li>• Imitation: spontaneously or provoked, meaningful or meaningless;</li> <li>• Eye gazing: provoked or spontaneously<sup>‡</sup>;</li> <li>• Pointing or alternative signals with referring function; and</li> <li>• Social smile: provoked or spontaneously.</li> </ul>	<p><b>Coping with changes (0.68/0.65):</b></p> <ul style="list-style-type: none"> <li>• Notice of changes: alertness or reaction rate; and</li> <li>• Responses to changes: acceptance or resistance.</li> </ul>	<p><b>Exploration and play (0.76/0.60):</b></p> <ul style="list-style-type: none"> <li>• Initiative to exploration;</li> <li>• Way of exploration;</li> <li>• Tactile defensiveness of materials;</li> <li>• Level of play; and</li> <li>• Variety of play: in number of objects and of different acts with an object.</li> </ul> <p><b>Problem-solving strategies (0.80/0.72):</b></p> <ul style="list-style-type: none"> <li>• Problem-solving behaviour; and</li> <li>• Asking for help: initiatives and quality.</li> </ul>

\* Internal consistency Cronbach's  $\alpha$  / intra-class correlation.

<sup>†</sup> Optional item only to be scored whenever a participant showed stereotyped behaviours with its own body of with objects that lasted 2 min or more.

<sup>‡</sup> Optional item for participants with enough residual vision to show eye gazing.

'eye gazing' were caused by the failure to observe eye gazing from the video recordings.

**Statistical analysis**

Non-parametric statistical methods were used, because variables were of ordinal level and the sample size was small. A rather liberal  $\alpha$  level of 10% was taken given the small number of participants. In case of multiple comparisons, the initial significance level was set at 1% to correct for chance capitalisation.

**Results**

The people with deafblindness and autism ( $n = 5$ ) were compared with the people with deafblindness without autism ( $n = 5$ ) with regard to the average total behaviour scores (see Table 3). Total behaviour scores ranged between 0 and 96. All participants gained scores far below the maximum score of 96 and far above the minimum score of 0. The participants with deafblindness and autism were rated significantly lower than the participants with

**Table 3** Total observation of characteristics of autism in persons with deafblindness (O-ADB) score\*

Deafblind with autism		Deafblind without autism	
Participant	O-ADB score	Participant	O-ADB score
1	47.75	6	70.00
2	45.50	7	77.25
3	49.50	8	75.75
4	53.50	9	64.25
5	39.00	10	71.00
Mean	47.05 (5.37)	Mean	71.65 (5.15)
(standard deviation)		(standard deviation)	

\* Excluding scores for 'eye gazing' and 'breaking stereotyped behaviour'.

deafblindness without autism (Mann-Whitney  $U = 0.000, P = 0.009$ ). This means that the people with deafblindness and autism showed qualitatively less target behaviours than the people with deafblindness without autism. Behaviours of both groups can be distinguished clearly from each

**Table 4** Comparison of observation of characteristics of autism in persons with deafblindness categories in deafblind participants with or without autism

Behavioural category	Mann–Whitney	P (one-tailed)	Persons with autism vs. without autism
Openness for contact	$U = 0.000$	0.005*	Less openness for contact
Reciprocity/joint attention	$U = 0.000$	0.007*	Less reciprocity
Communicative signals/functions	$U = 0.000$	0.005*	Less initiatives to contact and more instrumental/functional intention
Stereotyped behaviour	$U = 6.000$	0.162	More persistent stereotyped behaviour
Exploration and play	$U = 7.000$	0.125	Less variety in and less quality of explorative behaviour
Coping with changes	$U = 3.500$	0.03	More problems in coping with changes
Problem-solving strategies	$U = 4.000$	0.04	Less adequate strategies for problem-solving and less adequate questions for help

\* Significant for  $\alpha = 1\%$ .

other, because there is absolutely no overlap in the scores of individuals in both groups.

The main research goal was to establish differences in behaviours of people with deafblindness with or without autism in an unequivocal way. Therefore, the scores on the behavioural categories were also compared between both groups (see Table 4).

Participants with deafblindness without autism showed higher scores than participants with deafblindness and autism on all behavioural categories. Significant differences ( $P < 0.01$ ) between the people with deafblindness and autism and those without autism were found for 'openness for contact', 'reciprocity/joint attention' and 'communicative signals/functions'.

Nearly significant differences ( $0.01 < P < 0.05$ ) were found for 'coping with changes' and 'problem solving strategies'. The differences for 'stereotyped behaviour' and 'exploration and play' were not significant. Seven deafblind participants with or without autism showed some repetitive movements. Stereotyped behaviours of all participants except one stopped spontaneously within 2 min or after the examiner initiated social interaction. Individual variation in persistency and in the affect on exploration and social contact was found in both groups of participants and did not differentiate both groups. Exploration and play behaviours were restricted to manipulating objects. Only a few times, rudimentary forms of functional play were seen. In most cases, this was only direct imitation of the functional play shown by the psychologist.

As most participants with autism were male, gender but not autism could have explained the differences. We therefore analysed the variables from Table 4 again with gender as the independent variable. No significant differences were found. Note also that all individual scores in the autism group were lower than the lowest score in the non-autism group. On an individual basis, we compared the female participant with autism with the male participant in the autism group. She only had the highest score with regard to coping with changes but not, as might have been expected, on the social and communicative categories. The male participant in the non-autism group was also compared with the female participant in this group. His scores were not lower than the scores of her on any of the categories. For openness to contact and reciprocity, he even had the highest score. In conclusion, autism but not gender seems to be responsible for the differences in the O-ADB categories.

## Discussion

Up till now, empirical data on autism in deafblind persons with ID are not available. This study is the first to present empirical data by means of a semi-structured observation. The observation-instrument developed for this purpose (see Hoevenaars *et al.* 2008) seems to be useful to determine differences in behaviours of people with deafblindness and autism and of people with deafblindness without autism for the categories: 'openness for contact',

'reciprocity in contact including joint attention' and 'communicative functions'. For the categories 'coping with changes' and 'problem solving strategies', the results are not significant enough to conclude that both groups differ from each other. The categories 'stereotyped behaviour' and 'exploration and play' did not differentiate between the two groups of deafblind persons. In the case of stereotyped behaviour, this might be the result of a lack of reliability in observing these behaviours, which is odd because beforehand we expected these behaviours to be easy to observe.

Based on the literature and our results, we can state that people with deafblindness and autism show significantly more autism-specific behaviours than people with deafblindness without autism. We subsequently conclude that it is possible to differentiate deafblind people with autism from deafblind people without autism. However, there are more factors affecting developmental outcome in deafblind people with profound ID (Van Dijk & Nelson 1996; Freeman & Groenvelde 2000; Rødbroe 2001). For instance, sensory and social deprivation, brain damage and ID all affect a person's behaviour. These factors might explain the many similarities between people with deafblindness and autism and people with deafblindness without autism that were also found in this study. Given the fact that both groups did not differ in the level of ID, we can rule out ID as an alternative explanation for the differences in behaviour between deafblind intellectually disabled persons with or without autism. Anecdotal information does not point out any differences in sensory and social deprivation between both groups of deafblind persons. Brain damage might explain some of the variance, but it is unlikely that variance explained by brain damage is not already explained by level of ID.

With regard to the DSM-IV-TR criteria of autism, the current study found differences in behaviour of people with deafblindness and ID with or without autism in 'social interaction' and in 'communication'. No differences were found in 'restricted patterns of behaviour, interests and activities'. These results are in correspondence with results of studies on autism in persons with visual impairments. For these persons, similarities were found for the amount of stereotyped behaviours and the use of atypical language, but blind persons

with autism showed less quality of social interaction and communication in comparison with blind persons without autism (Brown *et al.* 1997; Hobson *et al.* 1997). At first glance, these results do not correspond to the similarities in the social impairments (Hobson & Bishop 2003) found between blind children with severe intellectual impairments and sighted children with autism. ID but not blindness *per se* provoke behaviours that look like autistic behaviours. If this is the case, all the participants in the current study should have been diagnosed as autistic. The difference with the Hobson and Bishop's study is that the authors attributed their findings to a diagnostic overshadowing bias. The diagnostic overshadowing bias describes the tendency of clinicians to overlook symptoms of mental health problems in this client group and attribute them to being part of 'having an intellectual disability' (Mason & Scior 2004). It seems that the presence of ID decreases the diagnostic importance of abnormal behaviour. In the present study, all participants had ID, so that diagnostic overshadowing with regard to ID cannot be the case. Deafblindness itself might add an extra overshadowing bias next to ID, leading to false positive diagnoses of autism in people who are deafblind. Our study showed that diagnoses of autism in people who are deafblind and intellectually disabled should be based on social interaction and communication skills.

Just as important as the differences are the similarities between deafblind people with ID with or without autism. No differences were found for exploration, play and problem-solving behaviours. Probably, the visual impairment sets a lower limit for the exploration and play behaviours. As Andrews & Wyver (2005) nicely reviewed, blind children tend to spend more time in solitary play, display play behaviours that are perseverative and, because of lack of vision, show minimal spontaneous imitation.

The case is less clear for stereotyped behaviours because they could not be observed reliably. Repetitive actions and stereotypic movements are a common component of mental disturbance and severe brain disorders (Andrews & Wyver 2005). It is also known that all congenitally blind children show one or more stereotyped behaviours (Jan *et al.* 1997; Tröster *et al.* 1991), and the prevalence is also

rather high (10–45%) in visually impaired children (Jan *et al.* 1977). Based on these statistics, stereotyped behaviours are not good candidates for differential assessment of autism in deafblindness because baseline levels are too high. However, stereotyped behaviours are interesting for the differential assessment because of differences in the underlying process. According to Gense & Gense (2005), initiatives for interaction could increase stereotyped behaviours in persons with autism, because of the possible stressful nature of the interaction. The function of stereotyped behaviours is thought to be to reduce these stress levels. However, in contrast to this theory, Van Dijk & Janssen (1993) and Murdoch (2000) proposed that stereotyped behaviours in people with deafblindness are caused by a lack of stimulation, information and communication. For certain behaviours such as body rocking, there is a third explanation, namely an attempt to seek physical proximity (Van Dijk & Janssen 1993). So, for people with deafblindness, stereotyped behaviours are expected to decrease or stop completely after social interaction is initiated. In the current study, stereotyped behaviours of all participants except one stopped spontaneously within 2 min or after the examiner initiated social interaction. These results seem to be in contradiction to the theory that stereotyped behaviours in persons with autism are resistant to intervention. However, in the current study, both the visual impairments and the ID might have caused stereotyped behaviours, whether the person is autistic or not. The similarities found in stereotyped behaviours in individuals with deafblindness and ID make stereotyped behaviours a poor characteristic for differential diagnosis of deafblindness and autism in persons with additional ID.

Level of play was not associated with the presence of autism in the deafblind participants. Probably, the severe ID caused the low levels of play. In typical children, functional play develops at the age of 12–18 months and symbolic play at the age of 18–24 months (Sigman & Ungerer 1981). The participants in the current study were all severely intellectually disabled, meaning that their mental ages were below the chronological age at which a typical developing child shows symbolic play and, in some cases, functional play. As a result, it is not the autism but the intellectual development that set a

limit to the level of play that was found. Note that this does not mean that, in deafblind people with moderate or no ID, exploration and play could not be good diagnostic behaviours for assessing autism.

One could fear that, by lack of suitable instruments, the experts diagnosed the deafblind participants mainly on the basis of social and communicative skills and that the results reflect the clinical selection process by finding differences with regard to social and communication skills. Note, however, that the panel consisting of a psychiatrist, psychologist and an independent expert in deafblindness all had ample experience with deafblind people. The general belief in deafblindness experts is that autism is over-diagnosed in deafblind people. So experts in deafblindness tend to disregard social and communicative behaviours in their own assessments given the large topographical overlap in overt behaviours but differing underlying mechanisms in deafblind and autistic people. We are therefore not afraid that the diagnosis of autism and, as a result, the selection of participants is mainly based on social and communicative behaviours, which in turn would explain the study results. In contrast, if there would have been a selection bias, we would have expected this to concern stereotyped behaviour, exploration and play as well as coping with changes. Note also that all items of the O-ADB, including the social and communicative items, take language and social problems of deafblind people into account. To conclude, we strongly believe that the results reflect differences between profoundly intellectually disabled deafblind people with or without autism and is not the result of a selection bias.

A problematic part of this study was the way we assessed autism in persons who are deafblind and intellectually disabled. There is no consensus about how to put the label autism on them, as we lack suitable diagnostic instruments to classify autism in the deafblind and as experts familiar with autism, deafblindness and ID are scarce. That is why a consensus judgment of three professionals each with different background was used. But with this procedure, it was still not possible to reach consensus for one-third of the institute's population. For these persons, the presence of autism remains uncertain. This makes it hard to offer these persons the best possible interventions for developing communica-

tion and language (Carvill 2001) and to support the development of cognitive concepts (Noens & Van Berckelaer-Onnes 2004).

Several other methodological problems affected this study. First, the study sample was rather small. This was the result of the strict inclusion rules. As a result, only a few deafblind people were eligible for this study. The small number of participants with heterogeneous aetiologies makes it hard to generalise the conclusions to the total population of deafblind people. From the studies on autism in blindness, we know that the aetiology of the visual impairment is probably not important, as autism does not seem to be related to any particular visual disorder but most likely to brain damage and developmental setback or delay (Andrews & Wyver 2005). Second, serious attempts were made by the examiners to follow the participants' interests and initiatives in order to achieve the best possible cooperation by the participants and thus the most valid results. However, the standardisation of the assessment procedure restricted the opportunities for optimal adjustment to the participant's behaviour. As a result, some possible differences between the two groups of deafblind participants might have been missed, because individual adjustment is often a prerequisite to get deafblind clients involved in social interaction. Last, psychometric properties of the assessment procedure other than internal consistency and inter-rater reliability are unknown. Our assessment procedure is therefore experimental and awaits replication. Currently, we are in the process of adapting the O-ADB to make it a reliable and valid assessment tool for diagnosing autism in deafblindness (Hoevenaars *et al.* 2008; Westland *et al.* unpublished data). By concentrating on the behaviours that differentiate between deafblind people with or without autism and on the behaviours that in combination belong to the DSM-IV-TR classification autism, we hope to be able to construct an assessment procedure that can help the clinician to adequately diagnose autism in the population of deafblind people with ID.

## References

- American Psychiatric Association (2000) *Diagnostic and Statistical Manual of Mental Disorders DSM-IV-TR*. American Psychiatric Association, Washington, DC.
- Andrews R. & Wyver S. (2005) Autistic tendencies: are there different pathways for blindness and autism spectrum disorder? *The British Journal of Visual Impairment* **23**, 52–7.
- Brown R., Hobson R. P. & Lee A. (1997) Are there 'Autistic-like' features in congenitally blind children? *Journal of Child Psychology and Psychiatry* **38**, 693–703.
- Carvill S. (2001) Sensory impairments, intellectual disability and psychiatry. *Journal of Intellectual Disability Research* **45**, 467–83.
- Cass H. (1998) Visual impairment and autism: current questions and future research. *Autism* **2**, 117–38.
- Dale N. (2005) Early signs of developmental setback and autism in infants with severe visual impairment. In: *Autism and Blindness: Research and Reflections* (ed. L. Pring), pp. 74–98. Whurr Publishers, London.
- De Bildt A., Sytema S., Kraijer D. & Minderaa R. (2005) Prevalence of pervasive developmental disorders in children and adolescents with mental retardation. *Journal of Child Psychology and Psychiatry* **46**, 275–86.
- Freeman R. D. & Groeneweld M. (2000) The behaviour of children and youth with visual and hearing disability. In: *Developmental Disability and Behaviour* (eds C. Gillberg & G. O'Brien), pp. 27–42. Mac Keith Press, London.
- Frith U. (2003) *Autism, Explaining the Enigma*. Blackwell Publishing, Malden, MA.
- Gense M. H. & Gense D. J. (2005) *Autism Spectrum Disorders and Visual Impairment*. American Foundation for the Blind Press, New York.
- Hartshorne T. M., Grialou T. L. & Parker K. R. (2005) Autistic-like behavior in CHARGE syndrome. *American Journal of Medical Genetics* **133A**, 257–61.
- Hobson R. P. & Bishop M. (2003) The pathogenesis of autism: insights from congenital blindness. *Philosophical Transactions of the Royal Society of London* **358**, 335–44.
- Hobson R. P., Brown R., Minter M. E. & Lee A. (1997) Autism' revisited: the case of congenital blindness. In: *Blindness and Psychological Development in Young Children* (eds V. Lewis & G. M. Collis), pp. 99–115. British Psychological Society, Leicester.
- Hoevenaars M. A. A. & Antonissen A. C. F. M. (2007) *Deafblindness and autism spectrum disorders: how to differentiate?* Paper presented at the Deafblind International World Conference Perth, Australia, September.
- Hoevenaars M. A. A., Antonissen A. C. F. M., Vervloed M. P. J. & Knoors H. E. T. (2008) *An instrument for observation of autism characteristics in persons with deafblindness, O-ADB*. Unpublished manuscript.
- Jan J. E., Freeman R. D. & Scott E. P. (1977) *Visual Impairment in Children and Adolescents*. Grune & Stratton, New York.

- Jordan R. (2005) Educational implications of autism and visual impairment. In: *Autism and Blindness: Research and Reflections* (ed. L. Pring), pp. 142–57. Whurr Publishers, London.
- Knors H. & Vervloed M. P. J. (2003) Educational programming for deaf children with multiple disabilities. In: *Oxford Handbook of Deaf Studies, Language and Education* (eds M. Marschark & P. E. Spencer), pp. 82–94. Oxford University Press, New York.
- Kraijer D. W. (2003) Stereotiep gedrag, autismespectrumstoornissen en enkele syndromen [Stereotyped behavior, autism spectrum disorders and several syndromes]. *Wetenschappelijk Tijdschrift Autisme* **2**, 24–39.
- Krug D., Arick J. A. & Alsmund P. J. (1980) *Autism Screening Instrument for Educational Planning (ASIEP)*. ASIEP Ed. Co, Portland, OR.
- Le Couteur A., Rutter M. & Lord C. (2003) *ADI-R Autism Diagnostic Interview – Revised*. Western Psychological Services, Los Angeles, CA.
- Lord C., Rutter M., DiLavore P. C. & Risi S. (2006) *Autism Diagnostic Observation Schedule-WPS (ADOS-WPS)*. Western Psychological Services, Los Angeles, CA.
- Mason J. & Scior K. (2004) Diagnostic overshadowing amongst clinicians working with people with intellectual disabilities in the UK. *Journal of Applied Research in Intellectual Disabilities* **17**, 85–90.
- Murdoch H. (2000) Repetitive behaviours in children with sensory impairments and multiple disabilities. *DBI Review* **26**, 7–11.
- Nelson C., Van Dijk J., McDonnell P. & Thompson K. (2002) A framework for understanding young children with severe multiple disabilities: the Van Dijk Approach to Assessment. *Research and Practice for Persons with Severe Disabilities* **27**, 97–111.
- Noens I. & Van Berckelaer-Onnes I. (2004) Making sense in a fragmentary world: communication in people with autism and learning disability. *Autism* **8**, 197–218.
- Noens I. L. J. (2004) *Pieces of the puzzle: sense-making and communication in autism spectrum disorders*. Doctoral Dissertation, Leiden University, Grafimedia, Leiden, the Netherlands.
- Rødbroe I. (2001) Discriminating congenital deafblindness from other disabilities. *NUD Bulletin* **1**, 23–5.
- Sigman M. & Ungerer J. A. (1981) Symbolic play and language comprehension in autistic children. *Journal of the American Academy of Child Psychiatry* **20**, 318–37.
- Smith T. M., Nichols S. L., Issekutz K. & Blake K. (2005) Behavioral profiles and symptoms of autism in CHARGE syndrome: preliminary Canadian epidemiological data. *American Journal of Medical Genetics* **133A**, 248–56.
- Tröster H., Brambring M. & Beelman A. (1991) Prevalence of situational causes of stereotyped behaviors in blind infants and preschoolers. *Journal of Abnormal Child Psychology* **19**, 569–90.
- Van Dijk J. P. M. (1982) *Rubella Handicapped Children: the effects of bi-lateral cataract and/or hearing impairment on behavior and learning*. Lisse, The Netherlands: Zwets & Zeitlinger.
- Van Dijk J. & Janssen M. (1993) Doofblinde kinderen [Deafblind children]. In: *Meervoudig gehandicapt: een zorg apart [Multiply Handicapped an Unusual Care]* (ed. H. Nakken), pp. 34–73. Lemniscaat, Rotterdam.
- Van Dijk J. P. M. (1995) *Congenitaal doofblinde kinderen en de chaostheorie [Congenital Deafblind Children and Chaos Theory]*. Institute for the Deaf (Viataal), Sint Michielsgestel.
- Van Dijk J. P. M. & Nelson C. (1996) Syndromes, behavior, and educational intervention. *Deafblind Perspectives* **4**, 1–25.
- Van Dijk J. P. M., Klomberg M. J. M. & Nelson C. (1993) *Strategies in Deafblind Education Based on Neurological Principles*. Institute for the Deaf (Viataal), Sint Michielsgestel.

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