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Robert Didden [a]; Hubert Korzilius [a]; Peter Sturmey [b]; Giulio E. Lancioni [c]; Leopold M. G. Curfs [d]
[a] Radboud University Nijmegen, The Netherlands
[b] Queens College and City University of New York,
[c] University of Bari, Italy
[d] University of Maastricht, The Netherlands

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Preference for water-related items in Angelman syndrome, Down syndrome and non-specific intellectual disability

ROBERT DIDDEN[1], HUBERT KORZILIUS[1], PETER STURMEY[2], GIULIO E. LANCIONI[3] & LEOPOLD M. G. CURFS[4]

[1]*Radboud University Nijmegen, The Netherlands,* [2]*Queens College and City University of New York, USA,* [3]*University of Bari, Italy, and* [4]*University of Maastricht, The Netherlands*

Abstract

Background Few case controlled studies have been published on the behavioural phenotype of Angelman syndrome (AS). Little is yet known about preferences in individuals with AS.

Method Preferences for water-related items and non-water-related items were assessed in 27 individuals with AS and two matched groups of participants, one with Down syndrome (DS) and the other with non-specific intellectual disabilities (NS), using an adapted Dutch version of the Choice Assessment Scale (Matson et al., 1999).

Results Individuals with AS showed a higher preference for water-related items compared to individuals in both comparison groups, substantiating previous reports which have suggested that individuals with AS are fascinated with water.

Conclusions Knowledge about preferences in individuals with AS, especially with regard to water-related objects and activities, is important in person-centred planning of daytime activities and training programs for this group.

Keywords: *Angelman syndrome, preference assessment, comparative study*

Introduction

Angelman syndrome (AS) is a genetic neurodevelopmental disorder characterised by severe intellectual disability, absence of speech, seizures, microcephaly, ataxic gait, inappropriate laughter, restlessness, and sleep problems (Clayton-Smith & Laan, 2003; Williams, 2005). In most cases, the clinical phenotype of AS can be confirmed by genetic testing. Four types of genetic abnormalities involving the absence or dysfunctioning of the normally active maternal allele at the 15q11-q13 region are known to cause Angelman syndrome: deletion (70–75%), paternal uniparental disomy (2–5%), imprinting defects (2–5%), and UBE3A and other presumed single gene mutations (20–25%) (Clayton-Smith & Laan, 2003). Some progress has been made in describing the behavioural phenotype of AS (see e.g., Williams, 2005), however it should be noted that only a few studies have used case controlled studies in their investigations (e.g., Berry, Leitner, Clarke, & Einfeld, 2005). Knowledge about the behavioural phenotype for AS is important for providing good care for individuals with this syndrome, however little is yet known about the preferences of this group. Anecdotal reports suggest that they prefer and are fascinated by objects and activities that are related to water (Berry et al., 2005; Clayton-Smith & Laan, 2003; Williams, 2005). A fascination with water is considered to be one of the associated clinical characteristics of AS (Williams, 2005). However some surveys have found this to be the case for only 68% of their sample (Clarke & Marston, 2000), which invites the question of whether or not fascination with water is indeed a characteristic of AS.

Knowledge of personal preferences is required for educational and behavioural interventions, as well as to facilitate choice-making (Lohrmann-O'Rourke & Browder, 1998). Researchers have developed both direct and indirect methods of preference assessment for individuals with intellectual disability (ID). In direct assessment, the therapist presents actual items or activities to an individual and then observes for approach or avoidance behaviours, such as exhibiting a positive facial expression or pushing the item away. Direct preference assessments have been developed for use in day-care centres for young children with developmental delays (e.g., Didden & de Moor, 2004) and for individuals with profound disabilities

Correspondence: Dr Robert Didden, Department of Special Education, Radboud University Nijmegen, PO Box 9104, Nijmegen 6500 HE, The Netherlands. E-mail: r.didden@pwo.ru.nl

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living in residential facilities (e.g., Green, Middleton, & Reid, 2000; Reid, Everson, & Green, 1999).

Although often effective, direct preference assessments may be limited because they are time-consuming, require specialised skills, and are only able to assess a limited range of items (Matson et al., 1999; Sturmey, Matson, & Lott, 2003). Many individuals with ID have limited communicative skills, making it difficult for them to make their preferences known by telling others. This can be addressed by implementing indirect methods of preference assessment, whereby parents, teachers and direct care staff are asked to act as proxies in reporting the individual's preferences.

Matson and colleagues (1999) developed the Choice Assessment Scale (CAS), a 60-item checklist designed to systematically assess items that may function as reinforcers for individuals with profound to severe ID. Informants were direct care staff working in a large residential facility. Factor analysis yielded four subscales: (i) Tangibles, (ii) Activities, (iii) Sensory, and (iv) Edibles. Good psychometric properties have been reported for the CAS.

Didden et al. (2006) adapted and translated the CAS into Dutch for a large sample of individuals with AS ($N=105$). They adapted the CAS in two ways. First, the authors added a 20-item subscale consisting of activities and objects that individuals may wish to avoid or escape from. Second, they added 13 items that referred to water. The adapted version of the CAS therefore contained 93 items and five subscales.

Didden et al. (2006) found that individuals with AS strongly preferred water-related items over non-water-related items. While this study was the first to investigate preferences in AS in an empirical way, an important limitation was the lack of a comparison group. In other words, it is still unclear whether the outcome of this study is unique to this diagnostic group, or if similar results may be expected for other diagnostic groups. Thus in the present study, the preferences of individuals with AS (as assessed by the adapted CAS) were compared to those of individuals with Down syndrome (DS), and of individuals with non-specific intellectual disabilities (NS). Specifically, we investigated whether individuals with AS showed a greater preference for water-related items over non-water-related items compared to individuals with DS or NS.

Method

Participants and procedure

First, a group of 27 individuals were randomly selected from Didden et al.'s (2006) study sample of individuals with AS (mean age=16.2 years; $SD=12.6$). Next, in order to recruit a comparison group of individuals with DS, we contacted the principal manager of the Dutch Down Syndrome Foundation, who sent a questionnaire and accompanying letter to parents who had a child with DS, asking them to complete and return the adapted Choice Assessment Scale. Staff members of a residential facility who had cared for individuals with DS for at least 6 months were also asked to complete the CAS. A total of 51 scales were completed for individuals with DS. From this sample, 27 individuals were matched to those with AS on gender, age (in years), level of ID, residential setting, and daytime residence. The mean age of this group was 16.2 years ($SD=12.8$). Finally, in order to recruit a second comparison group consisting of people with NS (i.e., with no known syndrome), two residential facilities and day-care centres were contacted, and another 27 individuals were matched to those with AS and DS on the same criteria. The mean age of this group was 16.1 years ($SD=12.9$). The parents and/or staff members of these individuals were also asked to complete and return the adapted CAS.

Informed consent was obtained from all participating parents and staff members. However to avoid bias in reporting as much as possible, they were not informed about the specific aim of the study. Instead, they were told that the aim was to explore the extent to which individuals like a relatively large number of objects and activities.

Demographic characteristics of the participants are described in Table 1. Most were children and adolescents with profound or severe ID. The majority lived at home. There were no statistically significant differences between the three diagnostic groups on gender ($\chi^2(2)=0.10$, $p=.95$), age in years ($F(2, 78)<1$), level of ID (Fisher's Exact Test, $p=.99$), living setting ($\chi^2(2)=0.00$, $p=1.00$), or daytime residence (Fisher's Exact Test, $p=.99$). Fisher's Exact Test was conducted where, due to low cell frequencies, the assumptions of the chi-square test could not be met. Fisher's Exact Test only produces a significance value and no formal test statistic (Fleiss, 1981).

Measure

Choice Assessment Scale. The original version of the Choice Assessment Scale (CAS) is a 60-item rating scale for the assessment of preferences and identification of potential reinforcers for individuals with profound to severe ID. Each item is rated on a 3-point scale in which "0" represents "Not at all",

Table 1. Demographic characteristics of participants

Variable	Diagnostic group		
	AS	DS	NS
Gender			
Male	41%	41%	44%
Female	59%	59%	56%
Level of ID			
Profound	26%	22%	19%
Severe	52%	48%	56%
Moderate	7%	8%	7%
Unknown[a]	15%	22%	18%
Residential setting			
Home	59%	59%	59%
Residential facility	41%	41%	41%
Daytime residence			
Home	11%	11%	14%
School/daycare	55%	48%	45%
Activity centre	30%	41%	37%
Elsewhere	4%	–	4%

Note. [a]No outcomes of tests or scales were available. All individuals were estimated to function in the profound to severe range of ID.

''1'' represents ''Some'', and ''2'' represents ''Very much''. The CAS consists of four subscales: Tangibles (12 items), Activities (13 items), Sensory (14 items), and Edibles (21 items). Matson et al. (1999) reported good inter-rater reliabilities and high internal consistencies (Cronbach alpha) for each subscale and for the total scale. Sturmey et al. (2003) replicated these findings.

For the purposes of this study, the CAS was translated into Dutch and adapted in two ways. First, a 20-item subscale was added consisting of activities and materials that individuals may wish to avoid or escape from. Scores on items in this Escape/Avoidance subscale may provide clues about what individuals tend to dislike rather than like. Preference assessment procedures seldom assess such items. Second, 13 items were added that referred to water. These items were added across the subscales: ''sound of water'', ''hands/feet in water'' (Sensory), ''balloon with water'', ''snow globe'', ''water pistol'', ''water wheel''(Tangibles), ''taking a bath/shower'', ''swimming in pool'', ''visit to fountain'', ''visit to pond'' (Activities), ''water'' (Edibles) and ''swimming pool'', ''pond'' (Escape/Avoidance).

The adapted version of the CAS therefore consisted of 93 items and five subscales, with each item rated on the same 3-point scale as the original CAS. There were 22 items on the Edibles subscale, 16 items on the Tangibles subscale, 17 items on the Activities subscale, 16 items on the Sensory subscale, and 22 items on the Escape/Avoidance subscale. Fifty graduate students in psychology who were familiar with ID and who were not informed about the aim of the study were asked to rate each item of the adapted CAS as being water-related or non-water-related. An item was defined as water-related if there was an agreement of at least 90% between raters. Items from the Edibles subscale were not included in this assessment as most of these items contain at least some water. In Didden et al.'s (2006) study, the internal consistency of the total scale (α=.94), as well as for each of the subscales (α ranging from .81 to .92), was good to high.

Statistical analyses

For each of the three groups, mean subscale scores for water-related and non-water-related items were computed. Both between-group and within-group analyses were performed. Differences between the groups were calculated using ANOVAs, and post-hoc analyses were conducted. Differences between means were calculated for each subscale and total scale for each group, using paired *t*-tests. Difference scores on the water-related and non-water-related items were also computed and analysed by means of planned contrasts between AS, DS and NS in GLM ANOVA.

Results

Between-group analyses

With diagnosis as the independent variable, results of the between-group analyses showed a significant effect for most subscales and for the total scale (see Table 2). Means for water-related items (W) were statistically higher than means for non-water-related items (NW) on all subscales and on the total scale for individuals with AS compared to individuals with DS and with NS, with the exception of the Escape/Avoidance subscale. Overall, individuals with AS showed a stronger preference for water-related items than individuals from both comparison groups.

Furthermore, Table 3 shows that the AS group had a significantly greater preference for water-related items over non-water-related items than individuals from both the DS and NS groups.

Within-group analyses

Paired *t*-tests between water-related and non-water-related items for each subscale and the total scale showed that individuals with AS scored higher on water-related items on the Tangibles (t (25)=3.28,

Table 2. Mean item scores for water-related and non-water-related items on each subscale, *F* and *p*-values, eta-squared and post-hoc analyses of the three diagnostic groups

Subscale and items	Diagnostic group	Mean item score	*SD*	*F* (*df*1, *df*2)	*p*	η^2	Post-hoc
Tangibles (NW)							
	AS	0.93	0.33	3.68 (2, 77)	.03	.09	AS>NS
	DS	0.67	0.49				
	NS	0.64	0.44				
Tangibles (W)							
	AS	1.32	0.69	17.31 (2, 74)	.00	.32	AS>DS
	DS	0.40	0.64				AS>NS
	NS	0.41	0.61				
Activities (NW)							
	AS	1.09	0.29	0.63 (2, 77)	.54	.02	
	DS	1.13	0.46				
	NS	1.00	0.51				
Activities (W)							
	AS	1.47	0.44	5.08 (2, 77)	.01	.12	AS>DS
	DS	1.09	0.51				AS>NS
	NS	1.06	0.60				
Sensory (NW)							
	AS	1.26	0.34	13.00 (2, 78)	.00	.25	AS>DS
	DS	0.81	0.44				AS>NS
	NS	0.71	0.47				
Sensory (W)							
	AS	1.33	0.63	16.51 (2, 77)	.00	.30	AS>DS
	DS	0.65	0.66				AS>NS
	NS	0.46	0.44				
Escape/Avoidance (NW)							
	AS	0.61	0.34	0.26 (2, 77)	.77	.01	
	DS	0.54	0.36				
	NS	0.56	0.43				
Escape/Avoidance (W)							
	AS	0.10	0.29	0.99 (2, 71)	.38	.03	
	DS	0.11	0.43				
	NS	0.27	0.61				
Total scale (NW)							
	AS	0.98	0.19	5.78 (2, 78)	.01	.13	AS>DS
	DS	0.79	0.31				AS>NS
	NS	0.73	0.32				
Total scale (W)							
	AS	1.09	0.40	19.50 (2, 78)	.00	.33	AS>DS
	DS	0.58	0.37				AS>NS
	NS	0.56	0.28				

Note. NW=non-water-related items; W=water-related items.

$p<.001$) and Activities (t (25)=5.70, $p<.001$) subscales. Individuals with AS scored lower on Escape/Avoidance of water-related items (t (23)=−6.05, $p<.001$), indicating a greater attraction to water-related items. There were no significant differences between water-related and non-water-related Sensory items and the total scale, although that test did approach significance (t (26)=1.18, $p=.08$).

For participants with DS, there were significant differences between water-related and non-water-related items on the Tangibles subscale (t (24)=−2.36, $p<.05$) and total scale (t (26)=−4.04, $p<.001$). Individuals with DS showed a lower preference for water-related items than for non-water-related items. There was also a significant difference for the Escape/Avoidance subscale (t (26)=−5.40, $p<.001$), showing less avoidance of

Table 3. Difference scores and planned contrasts for preference of water-related items compared to non-water-related items for individuals with AS compared to individuals with NS and DS

Subscale and items	Diagnostic group	Mean	*SD*	*F* (*df*1, *df*2)	*p*	η^2	Planned contrast
Tangibles							
	AS	0.39	0.60	19.29 (1, 74)	<.001	.21	AS>NS, DS
	DS		−0.22	0.47			
	NS		−0.24	0.66			
Activities							
	AS	0.38	0.34	10.51 (1, 77)	.001	.12	AS>NS, DS
	DS		−0.04	0.59			
	NS		0.05	0.49			
Sensory							
	AS	0.08	0.56	4.99 (1, 77)	.01	.06	AS>DS, NS
	DS		−0.17	0.55			
	NS		−0.25	0.48			
Escape/Avoidance							
	AS	−0.47	0.38	0.95 (1, 71)	.17	.01	
	DS		−0.40	0.39			
	NS		−0.33	0.57			
Total scale							
	AS	0.11	0.32	18.47 (1, 78)	<.001	.19	AS>DS, NS
	DS		−0.21	0.26			
	NS		−0.17	0.30			

Note. The difference score was calculated by subtracting the score on water-related items from the score on non-water-related items. Positive difference scores indicated a preference for water-related items. Negative difference scores indicated a preference for non-water-related items. For the Escape/Avoidance subscale, positive difference scores indicated *less avoidance of* (*i.e., more attraction to*) water-related items.

water-related items. No significant differences were found for the other subscales.

Individuals with NS had significantly lower mean scores on water-related items than on non-water-related items on the Sensory subscale (t (26)= −2.66, $p<.01$) and total scale (t (26)=−2.96, $p<.01$). There was also a significant difference for the Escape/Avoidance subscale (t (26)=−2.96, $p<.01$), showing less avoidance of water-related items. No significant differences were found for the other subscales.

Discussion

This case controlled study is the first to demonstrate that individuals with AS show a stronger preference for water-related items than individuals with DS or NS, confirming this aspect of the behavioural phenotype of AS. Within-group analyses for the comparison groups show either that water-related items were less preferred than non-water-related items, or that there were no significant differences in mean scores between the two types of items. Overall, results of the present study corroborate results found in previous studies and anecdotal reports that AS is associated with a fascination for water (e.g., Berry et al., 2005; Didden et al., 2006).

In the Didden et al. (2006) study, no significant associations were found between a strong preference for water-related items and demographic variables (e.g., genetic subtype, level of ID, age). The fascination remains to be explained. Our clinical experience and reports by parents and staff members suggest that individuals with AS become excited about performing activities and manipulating materials that are water-related. Anecdotally, for example, at a swimming-pool or pond the individual may become so excited that they might rush towards the pool in the absence of close supervision.

Within the group of individuals with AS, water-related items on the Activities and Tangibles subscales were the most highly preferred. Such activities and objects should therefore be incorporated into any educational or daytime activity program for individuals with AS. When designing person-centred plans for individuals with AS, they should be given access to water-related activities (e.g., visit to a fountain, taking a bath) and play materials (e.g., water wheel, balloon filled with water) as these items are highly preferred by these individuals. Furthermore, such items and activities may be used as reinforcers in educational programs, for example, during training in alternative communicative behaviours and adaptive skills (Duker,

Didden, & Sigafoos, 2004). However, strong preferences and reinforcers can be problematic. For example, Ishmael, Begleiter, and Butler (2002) reported on the case of a young child with AS who died by drowning in a wading-pool. They attributed the child's death to a fascination with water and water-related activities, and alerted parents and other caregivers to the danger posed by water for individuals with AS.

Rating scales such as the CAS can easily be incorporated into person-centred plans for identifying preferred items and activities (see Green et al., 2000). The CAS appears to be a suitable instrument for the assessment of preferences in individuals with profound and severe ID. Its psychometric properties are good to excellent, and it is sensitive to differences between subgroups of items (e.g., water-related and non-water-related items). Indirect preference assessment measures such as the CAS (and other rating scales) may be appropriate if the sample size of individuals and/or items is relatively large. However, it should be noted that although indirect methods of preference assessment are often used, they may not always be valid. Several studies have shown that there may be discrepancies between the outcome of direct and indirect assessment methods (e.g., Reid et al., 1999). What a caregiver believes the individual prefers or likes might differ from what the individual would select when presented with the actual item or activity. Items identified as preferred by proxies might not always be preferred by the individual, and might therefore not function successfully as reinforcers. Future studies should be conducted on the validity of the CAS to determine whether preferred items identified using the CAS would actually be chosen by individuals with AS (and others) during direct preference assessment, and whether these preferred items would function as reinforcers in educational and recreational programs.

Author note

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