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Preferences in individuals with Angelman syndrome assessed by a modified Choice Assessment Scale

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Abstract

Background Individuals with Angelman syndrome (AS) seem to have a strong preference for water-related items. Until present, preference assessment in AS has not been reported.

Methods An adapted Dutch version of the Choice Assessment Scale (CAS) was administered by parents and other caregivers to 105 individuals with AS. The CAS was adapted by adding several items related to water and by adding a sub-scale describing activities and materials that individuals may avoid or escape.

Results The five sub-scales and total scale of the modified CAS had good internal consistency. Water-related items were more often scored as preferred than non-water-related items. No associations were found between sub-scale and total scale scores and demographic characteristics (e.g. genetic subtype, age).

Conclusions This study shows that people with AS often have strong preferences for water-related items. The modified CAS is a reliable and sensitive instrument to assess client preferences.

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Introduction

Angelman syndrome (AS) is a neurodevelopmental disorder which is diagnosed clinically. The diagnosis can be confirmed by either cytogenetic or DNA testing in about 80–85% of clinically diagnosed individuals. Four different genetic mechanisms have been shown to give rise to AS: (1) deletions of region 15q11–13 of the maternally derived chromosome (70–75% of the cases); (2) paternal uniparental disomy of chromosome 15 (2–5% of the cases); (3) methylation imprinting mutations (2–5% of the cases); and (4) UBE3A and other presumed single gene mutations 20–25% of the cases) (Knoll *et al.* 1989; Buiting *et al.* 1995; Kishino *et al.* 1997; Matsuura *et al.* 1997; Ohta *et al.* 1999). Knowledge of the clinical phenotype of AS is important for providing good care for the affected individuals. Progress has been made with regard to describing the behavioural phenotype. Relatively many individuals with AS show distinct behavioural and developmental features such as inappropriate laughter, restlessness, communication deficits (Didden *et al.* 2004a), and sleep problems (Didden *et al.* 2004b). Individuals with AS typically function in the profound to low-moderate range of

intellectual disability (ID) and most have a seizure disorder (Clayton-Smith & Laan 2003).

Little is known about the preferences of individuals with AS. Several authors have noted that these individuals tend to be attracted to and fascinated with water. This is considered to be one of the associated clinical characteristics of AS (Williams 2001; Clayton-Smith & Laan 2003). Ishmael *et al.* (2002) reported that a 9.6-year-old child with AS died by drowning in a wading pool. They attributed the boy's death to a fascination with water and water-related activities. They alerted caregivers to the dangers raised by water in individuals with AS. Important as they may be, the conclusions are based on anecdotal observations. A systematic preference assessment in individuals with AS has not yet been published.

A preference assessment is a systematic procedure to identify an individual's preference for materials or activities. Items that an individual prefers may be used in designing programmes for training adaptive skills and treating problem behaviours, to facilitate choice making, and to enhance the individual's quality of life (Lohrmann-O'Rourke & Browder 1998). Both direct and indirect preference assessments have been used. In case of direct assessment, actual items are presented to the individual for a brief period and his or her response to this item is recorded. Although often effective, several limitations of this method have been identified. Direct assessment may be time-consuming and expensive, a limited range of items are assessed, items addressing social events are hard to assess directly and multiple sessions may be required because individuals with severe disabilities fatigue easily. Many staff and parents may find it difficult to apply such methods, although they may acquire these skills relatively quickly (Matson *et al.* 1999; Lavie & Sturmey 2002; Sturmey *et al.* 2003; Didden & de Moor 2004). Indirect assessment refers to procedures where parents and other caregivers are asked to act as proxies in reporting preferences of the individuals they care for. Such procedures may be used in a large sample of individuals and/or with a large range of items. No specialized training of parents and other caregivers is required. Thus, indirect assessment remains important in identifying preferences in individuals with severe disabilities.

Matson *et al.* (1999) developed the Choice Assessment Scale (CAS), a 60-item checklist. They used the CAS in 185 individuals with profound to severe ID to indirectly assess items that may function as reinforcers. Informants were direct care staff working in a large residential facility. Factor analysis yielded 4 sub-scales: Edibles, Activities, Sensory, and Tangibles. Matson *et al.* found that the CAS had high internal consistency and good test–retest reliability. In a sample of 206 individuals with severe to profound ID, Sturmey *et al.* (2003) also found high internal consistency for the CAS. Factor analysis yielded a four factor structure. These studies indicate that the CAS is a psychometrically sound instrument to identify preferred, potentially reinforcing activities and materials in individuals with severe and profound ID.

The aim of the present study was to assess the preferences in a relatively large sample of individuals with AS using the CAS. We investigated whether water-related items were more preferred than non-water-related items, and whether preferences for items were associated with demographic characteristics.

Materials and methods

Participants and procedure

The Dutch version of the adapted CAS (see below) was sent to 138 parents who were member of the Dutch Angelman Parent Association and who had a child with AS. In an accompanying letter, parent(s) were asked to complete this scale jointly. To control for a possible bias, parents and other caregivers were not informed about the aims of the study. In case the participant was living in a residential facility or in a community group home parents were asked to complete the questionnaire together with a staff member who knew the participant well for at least 6 months. If the CAS was not returned within 4 weeks, a reminder was sent to those parents.

The questionnaire was completed for 105 individuals, constituting a response rate of 76%. Their mean age in years was 15.5 (SD = 10.3; range: 1–45) and 55% were male. Of the participants, 74% lived at home, 22% lived in a residential facility, and 4% lived in a group home. The diagnosis of AS was based on chromosomal/DNA testing in 86% of the cases and 79% of this group had deletion, 11% had uniparental disomy, 5% had translocation or inversion, and 5% had UB3A gene mutation. Percentages of individuals

who had profound, severe, and moderate ID were 29, 45, and 6 respectively. In the remaining cases, level was unknown but was estimated to be in the profound to moderate range.

Choice Assessment Scale

The CAS is a 60-item reinforcer assessment scale. Each item is rated on a 3-point scale in which '0' represents 'not at all', '1' represents 'some', and '2' represents 'very much'. The CAS consists of four sub-scales: Edibles (21 Items), Tangibles (12 Items), Activities (13 items), and Sensory (14 items). Matson *et al.* (1999) reported good interrater reliabilities and high Cronbach alpha's for each of the sub-scales and the total scale. Sturmey *et al.* (2003) replicated these findings. We translated the CAS into Dutch and adapted the scale in two ways. First, a 20-item sub-scale was added addressing activities and materials individuals may avoid or escape from. Scores on items in this scale may provide clues about what individuals tend to dislike rather than like. Items an individual dislikes may function as negative reinforcers, for example, for problem behaviours. Procedures of indirect preference assessment seldom assess such items (for a review see Duker *et al.* 2004). Second, 12 items were added that referred to water (see Table 1). Fifty graduate students in psychology who were familiar with ID were asked to rate each item of the adapted CAS as being a water-related item or a non-water-related item. They were not informed about the aims of the study. An item was defined as water-related if there was agreement of at least 90% between raters. Items from the sub-scale Edibles were not included in this assessment as each item contains water.

The adapted version of the CAS contained 92 items and 5 sub-scales. Thus, there were 22 items on the Edibles scale, 16 items on the Tangibles scale, 16 items on the Activities scale, 16 items on the Sensory scale, 22 items on the new Escape/Avoidance scale and 12 new items related to water.

The mean item, total and sub-scale scores were computed. Values of Cronbach's alpha and item-total (minus item) point-biserial correlations were calculated for each sub-scale and the total score. The association between demographic variables with total and sub-scale scores were calculated using *t*-tests and ANOVA's as appropriate.

Results

The means and SD's for the sub-scales were: Tangibles – 11.48 (6.01); Escape/Avoidance – 12.21 (8.44); Edibles – 16.81 (10.37); Sensory – 16.91 (6.07), and Activities – 18.35 (5.70). Mean item scores are presented in Table 1. Results of paired *t*-tests revealed that differences between sub-scale mean item scores were all statistically significant, except for the difference between mean item scores of the sub-scales for Activities and Sensory and for Edibles and Tangibles (see Table 2).

The value of Cronbach's alpha for each of the sub-scales were 0.92 (Edibles), 0.83 (Tangibles), 0.81 (Activities), 0.82 (Sensory), and 0.87 (Escape/Avoidance). Cronbach's alpha for the total scale was 0.94. For each of the five scales and the total scale the median (and range) of item-total minus item point biserial correlations were: 0.58 (0.23–0.72), 0.44 (0.01–0.52), 0.37 (–0.10–0.53), .47 (0.00–0.74), .49 (0.16–0.61), 0.37 (0.00–0.92). Thus all five scales and the total scale were reasonably homogeneous.

Within each sub-scale and the total scale the mean item scores for water-related items were statistically significant higher than mean scores for other items (see Table 3). Within the Escape/Avoidance sub-scale water-related items had lower mean item scores than mean scores of other items.

There were no statistically significant relationships between mean sub-scales and total scale scores and the cause of AS (deletion vs. other causes), level of ID (moderate vs. severe vs. profound), gender and place of daytime activities (home vs. special school vs. daycare center). Significant differences between individuals with AS who lived in a residential facility and those who lived at home were found only for the Edibles sub-scale. Individuals who lived in a residential facility ($N = 23$; mean = 1.16) had a higher mean score on food- and drink-related items than those who lived at home ($N = 77$; mean = 0.93), $t[100] = 0.23$, $P < 0.05$. As foods and drinks are not readily available for individuals who live in a residential facility, this may account for the increased preference for these items in these individuals.

Finally, there were no statistically significant differences in mean score between water-related items and non-water-related items pertaining to each of the above demographic characteristics.

Table 1 Sub-scales, and mean scores of items of the adapted Choice Assessment Scale (CAS)

Sub-scale/Item (*N*)	Mean	Sub-scale/Item (*N*)	Mean	Sub-scale/Item (*N*)	Mean
Edibles		Music (94)	1.60	Taking a bath*† (97)	1.71
Apple sauce (97)	1.15	Quiet (no sound) (80)	0.32	Extra break (64)	0.30
Jell-O (96)	1.30	Bells (85)	1.14	Staying up late (79)	0.47
Pudding (89)	1.34	Cologne/parfum (62)	0.35	Swimming in pool*† (97)	1.65
Ice cream (96)	1.25	Vibrating tube/objects (77)	0.99	Playing games (85)	0.96
Milk shake (73)	0.92	Sound of water*† (76)	1.09	Interaction with peers (91)	0.99
Gum (64)	0.09	Hands/feet in water*† (90)	1.52	Interaction with staff (94)	1.45
Candy (87)	1.13			Receiving praise (93)	1.44
Cake (96)	1.11	*Tangibles*		Walking the grounds (96)	1.24
Snow cones (86)	1.10	Balloons (97)	1.43		
Chips (91)	1.02	Smooth balls (89)	0.87	*Escape/Avoidance**	
Popcorn (79)	0.65	Contoured balls (85)	0.89	Teaching tasks (50)	0.72
Crackers (87)	0.53	Hair brush (81)	0.40	Work tasks (54)	0.67
Fruit (98)	1.18	Music instruments (95)	1.32	Self-help tasks (61)	0.79
French fries (94)	1.09	Rattling objects (93)	1.39	Chores (59)	0.49
Hot dogs (84)	0.96	Small toys (87)	0.67	Other people (81)	0.27
Hamburgers (85)	0.93	Money (79)	0.16	Staff/teacher (77)	0.17
Soda (94)	0.93	Puzzles (86)	0.41	Peers (80)	0.30
Milk (93)	0.70	Paper (94)	1.06	Family members (84)	0.26
Chocolate milk (92)	0.70	Writing/coloring utensils (88)	0.45	Specific staff/teachers (75)	0.48
Juice (91)	0.89	Jewelry (81)	0.28	Specific peers (78)	0.55
Coffee (79)	0.39	Snow globe*† (70)	0.66	Noise (87)	0.86
Water* (92)	0.48	Water wheel*† (80)	1.04	Places where people scream/yell (89)	1.01
		Waterpistol*† (82)	0.84	Crowded places (91)	0.86
Sensory		Balloon with water*† (81)	1.51	Places that are loud (90)	0.99
Small lights (87)	1.11			Change in routine (81)	0.85
Reflections mirror ball (82)	1.18	*Activities*		Hot places (67)	0.57
Colored projections (83)	1.05	Trip to the park (93)	1.25	Cold places (64)	0.50
Pictures (94)	1.46	Trip to the canteen (78)	0.90	Specific smells (51)	0.35
Magazines/books (92)	1.34	Visit to fountain*† (73)	1.05	Specific food/tastes (83)	1.07
Looking out window (92)	1.01	Visit to pond*† (78)	0.90	Specific places (61)	0.66
Bubble tube/lava lamp (64)	0.75	Trip to community (97)	1.24	Swimming pool*† (87)	0.31
Mirror (92)	1.14	Bus rides (94)	1.47	Pond*† (69)	0.19
Television (99)	1.82	Dancing (87)	1.15		

* Item added to original CAS.

*† Water-related item.

Discussion

This is the first study in which a systematic, indirect preference assessment was conducted in individuals with AS. Persons with AS had stronger preferences for water-related than non-water-related items and were less likely to avoid water-related than non-water-related items. Thus, this study confirms the clinical impressions of earlier studies (Williams 2001; Ishmael *et al.* 2002; Clayton-Smith & Laan 2003) that AS is indeed characterized by a strong interest in water-related items. Their fascination with water remains to be explained. In order to strengthen the validity of our conclusions, comparative studies containing participants with other forms of ID, such as Down syndrome or Fragile X syndrome and non-specific forms of ID, should be conducted.

In general, activities and sensoric items were more preferred than foods and drinks and tangible items. Relatively high mean item scores were found for activities, such as, riding in a bus, taking a bath, swimming in a pool, interaction with staff and receiv-

Table 2 Differences between mean item sub-scale scores, *t* and *P*-values

Sub-scales	*N*	Mean	SD	t	d.f.	*P*
Edibles –	99	0.96	0.46	1.41	98	NS
Tangibles	99	0.89	0.44			
Edibles –	100	0.96	0.46	5.54	99	<0.001
Activities	100	1.18	0.37			
Edibles –	99	0.96	0.46	5.14	98	<0.001
Sensory	99	1.19	0.39			
Edibles –	94	0.94	0.46	4.74	93	<0.001
Escape/Avoidance	94	0.67	0.44			
Tangibles –	99	0.90	0.45	8.17	98	<0.001
Activities	99	1.19	0.37			
Tangibles –	99	0.90	0.45	7.90	98	<0.001
Sensory	99	1.19	0.39			
Tangibles –	94	0.89	0.45	3.80	93	<0.001
Escape/Avoidance	94	0.66	0.44			
Activities –	99	1.19	0.37	0.10	98	NS
Sensory	99	1.19	0.40			
Activities –	95	1.17	0.37	9.76	94	<0.001
Escape/Avoidance	95	0.67	0.44			
Sensory –	95	1.18	0.40	9.79	94	<0.001
Escape/Avoidance	95	0.67	0.44			

Table 3 Differences in mean scores between water-related and non-water-related items, *t* and *P*-values

Sub-scales	*N*	Mean	SD	t	d.f.	*P*
Tangibles NW –	95	0.84	0.41	3.81	94	<0.001
Tangibles W	95	1.08	0.77			
Activities NW –	99	1.13	0.38	6.20	98	<0.001
Activities W	99	1.39	0.51			
Sensory NW –	92	1.14	0.38	2.55	91	<0.01
Sensory W	92	1.31	0.68			
Escape/Avoidance NW –	87	0.66	0.42	6.37	86	<0.001
Escape/Avoidance W	87	0.25	0.58			
Total scale NW –	101	0.97	0.30	2.09	100	<0.05
Total scale W	101	1.05	0.48			

NW, non-water-related items; W, water-related items.

ing praise. These are activities that seem to be highly preferred by individuals with AS. Highly preferred items on the Sensory scale were watching television, listening to music, looking at pictures, and browsing through magazines and books. Balloons and manipulating musical instruments and rattling objects were highly preferred tangible items. Items with relatively high mean scores (see Table 1) may be incorporated into the educational and habilitative programmes for individuals with AS, and may be used as positive reinforcers during one-to-one training (see e.g. Duker *et al.* 2004).

Water-related items were highly preferred by individuals with AS. Excessively strong reinforcers can be problematic in that they may elicit approach behaviours that compete with other adaptive behaviours or may maintain maladaptive behaviours (Balsam & Bondy 1983) hence for some persons with AS man-

agement of water-related stimuli may require careful and individually based management.

The internal consistency of each sub-scale and the total scale of the modified CAS was good to high. The new scale to measure stimuli related to escape and avoidance, which might function as negative reinforcers, was also highly internally consistent. The new Escape/Avoidance scale had the lowest median of item-total minus item point biserial correlation (i.e. 0.37). Future research should investigate how to improve the sub-scale psychometrically (e.g. by developing better items, drop items from the scale).

The modified CAS was sensitive to the differences between water and non-water-related items in people with AS. This suggests that the modified CAS is a suitable instrument to assess client preferences in a number of different contexts. We did not conduct a factor analysis because criterion (i.e. subjects-to-variables ration should be no lower than 5; see Bryant & Yamold (1995) for conducting such an analysis was not met.

This study is the first study to empirically demonstrate a strong preference for water-related items in people with AS. However, it was limited in that it included only one sample of clients, but did not include other samples of clients with other forms of ID. Future research should investigate whether the preferred stimuli identified in the CAS function as reinforcers. Future research should also evaluate how indirect instruments, such as the CAS, can be incorporated into identifying stimuli that function as reinforcers in large-scale service settings.

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