

## **Behavioural and Psychological Symptoms of Dementia in Individuals With Down Syndrome**

*Emoke Jozsvai*

### **Abstract**

*This study compared the behavioural and psychological symptoms of dementia in Down syndrome (DS-D group) and people without history of mental retardation\* (NORM-D group). Non-compliance occurred at significantly higher rates in the DS-D than in the NORM-D group ( $\chi^2$  5.66,  $p < 0.03$ ). Non-compliance ( $F(1,62) = 6.85$ ,  $p < 0.01$ ) and property destruction ( $F(1,62) = 4.28$ ,  $p < 0.05$ ) were rated more severe for the DS-D than the NORM-D group. Insomnia was more severe among NORM-D group members than the DS-D group ( $F(1,62) = 67.57$ ,  $p < 0.00$ ). Management of dementia related challenging behaviours is discussed.*

Down syndrome (DS) (trisomy 21) is the most common recognized genetic condition associated with mental retardation\*, accounting for up to 22% of mental retardation cases with a known etiology (Murphy, Boyle, Schendel, Decoufle & Yeargin-Allsopp, 1998). Longevity for people with DS has increased almost sixfold over the last century. In the 1920's, the life expectancy for an individual with DS was 9 years, whereas today, the expected life span for a 1 year old child with DS is about 55 years (Thase, 1982; Strauss & Eyman, 1966). The oldest individual with DS reported in the literature was an 83 year old woman (Chicoine & McGuire, 1977). With the growth of the elderly DS population the number of individuals affected by dementia has increased. Dementia of the Alzheimer type is the most common form of dementia affecting adults with DS. Research suggests that probably all individuals with DS over the age of 35 develop the neuropathological changes characteristic of Alzheimer disease (Holland, Hon & Stevens, 2000), although not all these individuals will exhibit the clinical features of dementia. The clinical manifestations of dementia in DS increases with ageing from 8% in those between 35 and 40 years old to approximately 22% for those aged 40+, and approximately 60% in the 60+ age group (Lai & Williams 1989; Wisniewski, Silverman & Wegiel, 1984; Janicki & Dalton, 2000). Numerous physical conditions can diminish

cognitive functioning in DS and thereby mimic dementia. These include hypothyroidism, hearing loss, vision impairment, Vitamin B12 deficiency, sleep apnea, and depression (for a review of common medical conditions in DS see Cohen, 1999). It is important to identify these conditions because, once they are treated cognitive functioning often returns to its previous level.

The pattern of cognitive decline associated with dementia is similar in DS and the normative population (Oliver, Crayton, Holland, Hall & Bradbury, 1998), except that the decline in people with DS is from a below normal level of cognitive function. Affected members in both populations exhibit changes in orientation, progressive irreversible decline in memory, language skills, abstract thinking, motor skills, and the ability to perform activities of daily living (ADL). Associated symptoms of dementia are delusions, hallucinations, depression, activity disturbances (e.g., wandering, purposeless activity, and repetitive mannerisms), aggressiveness (e.g., verbal outbursts, physical threats, violence, and agitation), non-compliance (e.g., refusal to take medications, and perform hygiene routines), diurnal rhythm disturbances (e.g., insomnia), and affective lability (Zaudig, 1996). These behavioural changes are also seen in individuals with DS diagnosed with dementia (Moss & Patel, 1995; Prasher & Filler, 1995; Duggan, Lewis & Morgan, 1996; Cosgrave, Tyrrell, McCarron, Gill & Lawlor, 1999) and may pose a challenge for caregivers (Janicki, McCallion & 2000). Behavioural disturbances that are difficult to manage in the family home or in a typical housing situation for people with mental retardation may eventually result in a referral of the DS person to a generic extended care or nursing facility. Some long-term care facilities are reluctant to admit individuals with DS because they believe their behavioural symptoms of dementia are more problematic and more difficult to manage than that of demented elderly from the normative population. However, no data exist to support this assumption and there is a dearth of information that compares the behavioural and psychological symptoms of dementia in the two populations.

The aim of the present study was to evaluate whether or not the prevalence and magnitude of psychological and behavioural symptoms of dementia in DS are similar to those exhibited by individuals from the normative population with dementia. The Behavioral and Emotional Activities Manifested in Dementia (BEAM-D) Scale (Sinha et. al., 1992) was employed to gather information from primary caregivers about the prevalence and severity of problem behaviours and psychological symptoms of dementia in DS. Validity and reliability for the BEAM-D had been previously established by Dr. Sinha with a sample of 45 community dwelling, nursing home, or hospital residents (21 male, 24 female) without

a history of mental retardation and a primary diagnosis of dementia. The mean age of the subjects was 76.7 years ( $SD = 9.9$ ; range 58.4-96.6). As this sample represented a cross-section of demented elderly from the normative population, it was considered to be an appropriate comparison group for individuals with DS and dementia. Dr. Sinha kindly gave permission for an intergroup comparison to be carried out of his published data on demented people without mental retardation (the NORM-D group) with data prospectively collected on the individuals with DS and dementia (the DS-D group), the results of which are described below.

## Method

### Subjects

The DS-D group consisted of 19 people (10 male and 9 female) with DS and dementia (DS-D group). The mean age of the subjects was 53.9 ( $SD=5.7$ ) ranging from 42 to 62 years. The age difference between females and males was not significant ( $F= 0.78, p>0.05$ ). Fifteen individuals were residents in group homes and 3 were living in the community at the time of the evaluation. All subjects were diagnosed with dementia according to the DSM-IV criteria (American Psychological Association, 1994). They had at least one year of cognitive decline as measured by the Dementia Scale for Down Syndrome (DSDS) (Gedye, 1995). The DSDS is a 60-item caregiver interview instrument designed to assist clinicians diagnose dementia. The scale inquires about cognitive and behavioural signs characteristic of 'early', 'middle', and 'late' stages of dementia and yields a specific CCS (cognitive cut-off score) value for each stage. The DSDS also allows for a differential diagnosis of dementia with hearing and visual impairment, thyroid dysfunction, and depression. None of the subjects had a prior diagnosis of a major psychiatric disorder or uncorrected thyroid dysfunction. Three of the subjects were on neuroleptic medication at the time of the study. All subjects were classified with 'middle' stage dementia according to their CCS score on the DSDS. All the subjects were clients at Surrey Place Centre (Toronto, Canada), a community treatment facility for people with developmental disabilities.

### Procedure

The BEAM-D scale was administered to the primary residential support workers or family caregivers of the subjects by the author. Data collection was implemented in the context of regular clinical follow-up assessment

service. Caregivers were asked to rate whether or specific behaviours occurred during a 1-week interval that preceded data collection. The BEAM-D scale contains the following items:

*Target Behaviours*

1. Hostility/aggression: This item rates verbal and non-verbal expression of anger, resentment, and the presence of overtly aggressive behaviour (e.g. hitting, kicking or slapping others).
2. Destruction of Property (belonging to Self or Others): Wilfully damaging or destroying property, including tearing books, magazines, stuffing toilets with papers, and breaking items.
3. Disruption of Others' Activities: This item collects information on behaviours that interfere with or disrupt the activities of others (e.g., during social and group activities or activities of daily living) and are annoying or discomforting to others.
4. Uncooperativeness: This item gathers information on the extent to which the ratee is concerned about the needs and feelings of others, his/her co-operative behaviour and courtesy.
5. Noncompliance: This item gathers information about the extent to which the ratee is responsive to caregiver instructions, requests, or redirection.
6. Attention-Seeking Behaviour: These item rates behaviours exhibited to draw attention to the ratee and increase the demand on caregivers.
7. Sexually Inappropriate Behaviour: This item collects information on the extent to which the ratee engages in inappropriate sexual behaviour.
8. Wandering: This item rates persistent attempts to pass through locked doors and/or the extent to which the ratee wanders from designated areas.
9. Hoarding Behaviour: This item measures the extent to which the ratee collects and hides objects.

*Inferred States*

1. Depression: This item gathers information about the presence and severity of dysphoric mood and loss of interest or pleasure in activities.

2. Delusions: This item measures preoccupation with and belief in ideas that are not true and the effect of such beliefs on the ratee's behaviour.
3. Hallucinations: This item collects information about the frequency and degree of visual, auditory, and tactile hallucinations.
4. Anxiety: This item is based on the ratee's behaviour and/or verbal report. It assesses the individual's restlessness, worry or apprehension, fear and panic.
5. Appropriateness/Stability of Affect: This item rates the appropriateness, and lability of the ratee's affect.
6. Increased/Decreased Appetite: Changes in appetite are rated relative to the person's normal appetite or previously obtained baseline.
7. Sleep: This item gathers information about sleep disturbances that includes difficulty or inability to fall asleep, restlessness, and/or walking during the night.

Both target and inferred behaviours are rated on a 4-point scale. A score of 0 was given if the behaviour could not be assessed. A rating of 1 corresponds to the absence of the behaviour whereas ratings 2, 3, and 4 refer to the severity and/or intensity of the behaviour. The exception to this rating method is item Sleep that taps 3 distinct types, each on a 3-point scale.

Inter-rater reliability for the DS-D group could not be obtained for the BEAM-D scale because the majority of subjects had only one primary support worker reliably familiar with his/her behaviour.

## Results

Table 1 contains the percent frequency of target behaviours and inferred psychological states reported by caregivers and the result of chi-square tests comparing prevalence rates of the DS-D group and the normative group with dementia (NORM-D group). The Student's t statistic was employed to test the difference between the mean severity ratings of the observed and inferred behaviours of the DS-D group and NORM-D group. Table 2 displays the mean severity ratings of observable behaviours and inferred states for the two groups together with the results of the t tests.

Table 1 Frequency of behavioural problems reported on the Beam-D scale and results of chi-square test

BEAM-D Items	Norm-D DS-D (n=45) (n=15)		$\chi^2$	p
	f	f		
<i>Target Behaviours</i>				
1. Hostility/Aggression	35	52	1.62	0.27
2. Destruction of Property	8	26	0.13	0.75
3. Disruption of Others' Activities	22	36	1.47	0.23
4. Uncooperativeness	35	36	0.01	1.00
5. Noncompliance	27	57	5.66	0.03*
6. Attention-Seeking Behaviour	42	31	1.25	0.41
7. Sexually Inappropriate Behaviour	4	10	0.84	0.58
8. Wandering	29	42	1.06	0.39
9. Hoarding Behaviour	49	47	0.12	1.00
<i>Inferred States</i>				
1. Depression	58	52	0.78	0.41
2. Delusions	38	31	0.22	0.78
3. Hallucinations	29	47	1.39	0.27
4. Anxiety	71	57	1.21	0.38
5. Appropriateness/Stability of Affect	42	52	0.36	0.59
6. Increased/Decreased Appetite	29	26	0.15	0.77

\* Statistically significant difference

With the exception of attention-seeking, all target behaviours occurred at a higher frequency in the DS-D group than in the NORM-D group. However, only non-compliance occurred at a significantly higher rate ( $\chi^2 66, p < 0.03$ ) in the DS-D than in the NORM-D group. Sexually inappropriate behaviours were reported least frequently for both groups, whereas hoarding and uncooperativeness occurred at almost the same rates in the two groups. The inferred states of depression, delusion, anxiety, appetite changes, and insomnia occurred at higher frequency for the NORM-D than DS-D group, whereas the pattern of results for inappropriateness of affect and hallucinations was the opposite. The group differences in the frequency of inferred states were not statistically significant. However, the inferred state of insomnia (item Sleep) was rated significantly more severe for members of the NORM-D group ( $F(1,62)=67.53, p < 0.00$ ) than for the DS-D subjects. In contrast, property destruction ( $F(1,62)=4.28, p < 0.05$ ) and noncompliance ( $F(1,62)=6.85, p < 0.01$ ) were rated more severe by the support workers of the DS-D group than by caregivers of the NORM-D group.

Table 2. Severity ratings (means and standard deviations) of Beam-D Scale items and result of oneway ANOVA

BEAM - D Items	NORM-D (n = 45)		DS - D (n = 19)		F(1,62)	p
	M	SD	M	SD		
<i>Target Behaviours</i>						
1. Hostility/Aggression	1.53	0.81	1.68	0.75	0.47	0.49
2. Destruction of Property	1.15	0.52	1.52	0.90	4.28	0.04*
3. Disruption of Others' Activities	1.49	1.01	1.73	1.14	0.69	0.41
4. Uncooperativeness	1.55	0.84	1.63	0.95	0.11	0.74
5. Noncompliance	1.44	0.81	2.10	1.15	6.85	0.01*
6. Attention-Seeking Behaviour	1.80	1.06	1.45	0.96	1.53	0.22
7. Sexually Inappropriate Behaviour	1.11	0.53	1.10	0.31	0.01	0.94
8. Wandering	1.60	1.12	1.84	1.10	0.62	0.94
9. Hoarding Behaviour	1.95	1.17	1.84	1.01	1.28	0.72
<i>Inferred States</i>						
1. Depression	1.78	0.79	2.00	1.10	0.81	0.37
2. Delusion	1.84	1.19	1.36	0.83	2.55	0.12
3. Hallucinations	1.35	0.60	1.63	1.10	1.72	0.20
4. Anxiety	2.27	1.07	1.94	0.97	1.34	0.25
5. Appropriateness	1.49	0.63	1.78	0.85	2.28	0.14
6. Increased/Decreased Appetite	1.40	0.75	1.47	0.96	0.09	0.76
7. Insomnia	3.60	1.07	1.31	0.88	67.53	0.00

\* Statistically significant difference

## Discussion

This study compared dementia related challenging behaviours in DS and people without history of mental retardation. With the exception of noncompliance and property destruction, which occurred with a significantly higher frequency and magnitude in DS, the challenging behaviours of normative sample and the DS group were similar. Thus, people with DS and dementia could be cared for in regular long-term care or nursing home facilities.

Statistically significant group differences in noncompliance may be attributed to more severe comprehension deficits among members of the DS than in the normative group. Lack of compliance by DS individuals, who

may have a life-long history of relying on directions and following instructions, is likely to be rated more severe by caregivers than noncompliance by individuals without a previous history of mental retardation. Compliance with ADL (activities of daily living) can be enhanced by behaviour management techniques. This may include increasing and/or augmenting visual and verbal prompts, restructuring the environment to facilitate desired behaviours to occur, and providing enhanced physical assistance as dementia progresses and cognitive capabilities decline (Antonangeli, 1995).

Regarding property destruction, caregivers of individuals with DS reported repetitive paper tearing, stuffing tissue into toilet balls, and flushing hearing aids down the toilet. Common triggers for repetitive behaviours included under-stimulating environments and, fear or anxiety and/or misinterpretation of environmental stimuli. Environmental restructuring (i.e., removing objects and valuables) and increasing supervision may be effective to reduce property destruction. Repetitive behaviours such as tearing paper may be adaptive in virtue of reducing anxiety. Therefore, location and material for engaging in this behaviour may be designated within the environment.

Insomnia was the only inferred state that was rated more severe for the normative group than for members of the DS group. Providing consistent routines for walking, supervised physical exercise, limiting fluids containing caffeine or large amounts of concentrated sugars can reduce insomnia. Sleep pattern disturbances may also be reduced by eliminating triggers for nighttime fearfulness. Behaviour management and pharmacological support can help maintain or improve the quality of life for demented individuals and their caregivers. However, caregivers must consider whether the side-effects of a potentially helpful drug may be worse than the overall problem created by the behaviour.

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**Correspondence**

Emoke Jozsvai  
Surrey Place Centre  
2 Surrey Place  
Toronto, On  
M5S 2C2

[emoke@rogers.com](mailto:emoke@rogers.com)