Behavioural excesses and deficits associated with dementia in adults who have Down syndrome.

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Dementia in adults who have Down syndrome

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Abstract

Background. Informant based assessment of behavioural change and difference in dementia in Down syndrome can aid diagnosis and inform service delivery. To date few studies have examined the impact of different types of behavioural change.

Methods. The Assessment for Adults with Developmental Disabilities (A.A.D.S.), developed for this study, assesses behavioural excesses (11 items) and deficits (17 items) associated with dementia. Inter-informant reliability, internal consistency and concurrent validity were evaluated and found to be robust.

Results. A comparison of the AADS subscale scores for three groups (n = 12) of adults with Down syndrome demonstrated more frequent deficits and excesses and greater management difficulty and effects on the individual in a dementia group than age comparable and younger groups.

Conclusion. The AADS is a promising dementia specific measure for persons with intellectual disability. Further research should evaluate change as dementia progresses and the nature of management difficulty and effects on the individual.
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**Introduction**

In the last two decades there has been a significant increase in clinical and research interest in the assessment of dementia in people with intellectual disability. This increase is in part driven by the enhanced life expectancy of people with intellectual disability and the corresponding rise in the number of people who have Down syndrome who are surviving into their fifth decade and consequently at high risk for developing dementia (Eyman, Call and White, 1991). Holland, Hon, Huppert, Stevens and Watson (1998) have estimated the prevalence of dementia in people with Down syndrome to be 3.4% in the age band of 30 to 39 years, 10.3% in the age band of 40 to 49 years and 40% in those aged between 50 and 59. There is now a wealth of neuropathological, neuropsychological and related evidence that individuals who have Down syndrome are at high risk for developing Alzheimer’s disease (Oliver and Holland, 1986; Berg, Karlinsky and Holland, 1993) and there is a clear need for psychometrically robust assessments for research and clinical purposes.

Early descriptions of dementia in individuals with Down syndrome were largely anecdotal and focused primarily on emerging skill deficits and behavioural excesses (Dalton, Crapper and Schlotterer, 1974; Ellis, McCulloch & Corley, 1974; Haberland, 1969; Owens, Dawson and Losin, 1971; Ropper & Williams, 1980;). In the late 1970’s and early 1980’s cross-sectional age group studies employed neuropsychological assessments to assess the earlier signs of memory loss and other cognitive deficits (e.g. Thase, Tigner, Smeltzer & Liss, 1984; Wisniewski, Howe, Gwyn-Williams & Wisniewski, 1978). This early research was hampered by the paucity of appropriate test instruments for individuals who had intellectual disability.
and began to highlight the floor effects in cognitive tests and thus a potential role for informant based measures. More recent research has identified changes in behaviour and personality that are also observed in frontotemporal dementia (Ball et al., 2008; Holland et al., 1998; 2000, Nelson et al., 1995; 2001; Adams et al., 2010) and that are related to referral for intervention (Adams et al., 2008).

The two most significant problems for the assessment of specific neuropsychological deficits associated with dementia are the variability of degree of intellectual disability and the problems associated with administering neuropsychological tests to those with severe or profound intellectual disability who may not understand verbal instructions (Aylward et al., 1997; Crayton & Oliver, 1993; Crayton, Oliver, Holland, Hall & Bradbury, 1998; Oliver, 1999). In combination with the likely scenario that baseline measures are not available when an individual first presents with change that might indicate dementia, this issues have also led to reliance on informants and affirms a prevailing need for psychometrically robust informant based measures.

In longitudinal studies published to date, there is strong evidence that the neuropsychological impairments experienced by individuals with Down syndrome who develop dementia of the Alzheimer type are similar to those seen in the general population and there is a similar sequence of decline (Burt, Loveland, Chen, Chuang, Lewis & Cherry, 1995; Devenny, Silverman, Hill, Jenkins, Sersen & Wisniewski, 1996; Lai & Williams, 1989; Oliver, Crayton, Holland, Hall & Bradbury, 1998; Ball et al., 2008; Adams et al., 2010). There is also evidence of behavioural change and difference in longitudinal and cross-sectional studies and descriptions of clinical cohorts (Prasher and Filer, 1995; Dalton and Wisniewski, 1990; Oliver, Crayton,
Holland and Hall, 2000). As dementia progresses, neuropsychological assessment alone becomes less sensitive to change and behavioural signs become more prominent and clinically significant. It is clear therefore, that both neuropsychological and behavioural aspects of dementia warrant consideration in any assessment strategy.

At present there are few measures available that have been developed specifically for those who have intellectual disability. Two measures, the Dementia Questionnaire for Mentally Retarded Persons (DMR; Evenhuis, Kengen and Eurlings, 1990) and the Dementia Scale for Down Syndrome (DSDS; Gedye, 1995), have good face validity with some assessment of reliability and could be used alongside neuropsychological assessments. The need for caution when using these informant based measures is primarily related to the appraisal of inter-informant reliability. Given that there is high probability of change of carer over time the issue of inter-informant reliability is, arguably, more important than that of the short term stability. Diagnostic validity of these measures when used at one time point and alone is also an important issue. Prasher (1997) has suggested a modified scoring procedure for the DMR in order to overcome the potential problem of similarity of presentation of dementia and severe intellectual disability.

The need for dementia specific informant based assessments is also becoming more evident as service agendas evolve. Assessment is essential to clinical diagnosis and change over time is a more critical diagnostic issue for people with intellectual disability than the general population (Oliver, 1999). Clinically there is a need to address both the problems associated with diagnosis and proactive service planning and provision. There is good evidence that dementia in individuals with Down
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syndrome is associated with difficulties for carers and increasingly limited life experiences for the individual themselves (Oliver, Crayton, Holland and Hall, 2000; Adams et al., 2008). These clinical service response issues can be examined with appropriate measures that can identify behavioural difference associated with dementia and how this might impact on both carers and the individual.

The first aim of this study was to develop a reliable measure of the presence and frequency of behavioural deficits (the loss of adaptive behaviours such as the ability to recall information, practice self-care, show awareness of time, person and place) and excesses (the increase in frequency of behaviours to a point at which they interfere with daily life e.g. shouting, aggression) that are associated with dementia in persons with Down syndrome. Behavioural and psychological signs and symptoms of dementia are integral elements of the disease process (American Psychiatric Association 1994, Aylward et al 1997, Deimling & Bass 1986, Moss & Patel 1997, World Health Organisation 1992). There are many ways in which these can be grouped, for example by function e.g. sleep disorders, by altered behaviours e.g. wandering or by psychopathological symptom clusters e.g. behaviours indicative of memory loss. Another method of grouping would be behavioural excesses and deficits as the impact of these on carers and the individual might differ. This approach was adopted in this study. A second aim was to examine the degree of management difficulty and the effect on the individual that are associated with behavioural deficits and excesses experienced by persons with Down syndrome who have with dementia.

The design of the study incorporated a comparison of three groups of adults with Down syndrome (older adults with dementia, older adults with no dementia, younger
Dementia in adults who have Down syndrome adults with no dementia) in order to elucidate differences in behavioural excesses and deficits associated with dementia. The comparison of the two older adult groups was designed to identify dementia related differences whilst the comparison of the no dementia older and younger adult groups would identify purely age related differences. Prior to these analyses psychological assessments of the Dementia group and the other two groups were compared to evaluate integrity of the group allocation and the concurrent validity of the AADS was appraised by correlation of scores with established measures. A secondary analysis was employed to evaluate the impact of degree of intellectual disability on scoring on the informant measures.

**Method**

**Participants**

36 adults, 19 (52.8%) female and 17 (47.2%) male, with Down’s syndrome aged between 30 and 64 (mean = 45.17, s.d. = 9.36) were selected to take part in this study from a pool of 68 participants who have been enrolled onto a longitudinal study. Twelve participants were initially selected from the pool of 68 as they had been diagnosed with dementia by a psychiatrist or the psychiatrist determined there was probable dementia based on clinical examination and interview with carers about past and current functioning. This Dementia Group comprised five males and seven females (mean age = 49.0, range = 39 to 64, s.d. = 7.37). An Older Comparison Group (n =12), comprising participants aged over 40 who did not have a diagnosis of dementia nor was dementia suspected, was selected to be comparable to the dementia group on the basis of age (+/- 4 years) and gender (four males, eight females; mean
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age = 51.58, range 42 to 63, s.d. = 5.71). A Younger Comparison Group (n = 12) comprised participants aged 40 and under who did not have a diagnosis of dementia nor was dementia suspected (eight males, four females; mean age = 34.9, range 30.0 to 40.0, s.d. = 4.03). All participants were living in community settings, either small group homes or with their family. No participants had additional psychiatric diagnoses and there were no exclusion criteria with regard to behaviour.

Measures

**Neuropsychological Assessment.** The battery of neuropsychological assessments employed in cross-sectional and longitudinal studies by Crayton, Oliver, Holland, Bradbury & Hall (1998) and Oliver, Crayton, Holland & Hall (1998) was employed. This battery comprises seven sub-scales assessing: picture naming and identification, actions on request, orientation, object and picture memory and memory for sentences.

**Receptive language.** The British Picture Vocabulary Scales ‘ B.P.V.S.’(Dunn, Dunn, Whetton & Pintilie, 1992) was administered.

**Dementia.** The Dementia Questionnaire for Mentally Retarded Persons – ‘D.M.R.’ (Evenhuis, 1990; Evenhuis, Kengen & Eurlings, 1992) was completed by carers. This scale has two sub-scales: social behaviours and cognitive behaviours.

**Adaptive behaviour.** The Vineland Adaptive Behavior Scale ‘V.A.B.S.’ (Sparrow, Balla & Cicchetti, 1990) was completed with carers. There are three domains: Daily Living Skills, Communication and Socialization.
Assessment for Adults with Developmental Disabilities (A.A.D.S.) The A.A.D.S. is an informant based questionnaire developed for this study. Items were based on definitions of behavioural excesses and deficits associated with dementia outlined by DSM-IV (American Psychiatric Association, 1994) and the recommendations for evaluation procedures for dementia in persons with intellectual disability compiled by Aylward, Burt, Thorpe, Lai and Dalton (1997) to derive descriptions of observable behaviours related to such definitions. The questionnaire comprises twenty-eight items each rated on a seven point Likert scale. The questionnaire has two sub-scales that assess behavioural excesses (11 items: restlessness, night and day wandering, vocally disruptive, taking items, crying, uncooperative, verbally and physically aggressive, sexually inappropriate and repetitive speech) and behavioural deficits (17 items: inactivity, word finding difficulties, lack of interest, withdrawn, disoriented in time, person and place, becoming lost, two areas of self help, difficulties carrying out sequential acts, poor concentration, not alert, slow and falling) commonly associated with dementia. The Frequency of each item in the preceding two weeks is rated by the informant (‘More than once an hour/continually’, score 6, to ‘Has not occurred’, score 0). If an item is identified as having occurred at least ‘Once in the last two weeks’, score 1, a rating is then made to appraise Management Difficulty (‘no difficulty’, score 0, to ‘extremely severe difficulty’, score 6) and the Effect of the behaviour on the person who is showing the behaviour (‘no effect’, score 0, to ‘extremely severe effect’, score 6). Each point on the Likert scales is operationally defined. Total scores can be derived for Frequency, Management Difficulty and Effect for Deficits (maximum possible score for each is 102) and Frequency, Management Difficulty and Effect for Excesses (maximum possible score for each is 66). The
number of excesses and deficits can be calculated by counting the number of items scoring at least 1 on the *Frequency* scales.

To assess the inter-informant reliability and internal consistency the AADS was completed by carers of 49 participants with Down syndrome drawn from the pool of 68 participants enrolled onto the longitudinal study. Of these 49 participants, 29 (59.2%) were male and 20 (40.8%) were female. The mean age was 46.55 years (range 31 to 65, s.d. 9.42) and mean scores on the DMR cognitive and Social subscales were 15.0 (s.d. 12.23) and 14. 41 (s.d. 9.85) respectively. Eleven (22.4%) participants had been diagnosed with dementia or there was strong evidence for dementia. Of the 49 participants 22 (44.9%) participated in the group comparison study. Inter informant reliability was assessed by requiring two informants to independently complete the AADS within seven days of each other without conferring. Inter informant reliability indices, using Pearson product moment correlation coefficients, for the frequency of excesses and deficits and the number of excesses and deficits were: .81, .80, .86 and .80 respectively. For the management of excesses and deficits and the effect of excesses and deficits the indices were: .76, .83, .59 and .76 respectively. Thus, for the majority of the scales the inter informant reliability is good but the reliability index for the effects of deficits on the individual means that the data from this scale should be treated more cautiously. Assessment of the internal consistency of the frequency of excesses and deficits subscales yielded Cronbach’s Alpha coefficients of .79 and .89 respectively.

**Results**
Prior to analysis of the frequency and number of behavioural deficits and excesses associated with dementia and the impact of these behaviours the mean ages and gender breakdown of the groups were compared. The mean ages of groups were significantly different ($F(2, 33) = 28.05, p<.001$) with Scheffe’s post hoc analysis showing a significant difference between the Younger Comparison Group and both the Older Comparison Group and the Dementia Group but no difference between the Older Comparison Group and the Dementia Group. There was no difference between the groups in terms of gender.

Initial analyses examined the integrity of allocation to groups by comparison of the results of the D.M.R. and neuropsychological assessments. The group mean scores on these assessments and the results of statistical comparisons are shown in Table 1.

The data presented in Table 1 show a number of significant differences between the groups on assessments. The Dementia Group scored significantly higher than the other two groups on both subscales of the DMR and tests of orientation, and object and picture memory. Additionally, the Dementia Group scored significantly lower than the Older Comparison Group on performing actions on request and significantly lower than the Younger Comparison Group on Memory for Sentences (1). Overall the results of these comparisons demonstrate integrity to allocation to the Dementia Group with supportive evidence from both informant based and neuropsychological assessments.
To examine the concurrent validity of the Frequency subscales for Deficits and Excesses, the total Frequency scores were correlated with D.M.R. sub-scales across all groups. Additionally, an analysis was undertaken to examine the association between the AADS Frequency scores, D.M.R. scores and degree of intellectual disability in the non demented groups to evaluate whether the dementia measures were influenced by degree of intellectual disability in those who are not showing dementia. Consequently correlations were also calculated for the total Frequency score of the AADS, the DMR scores and the BPVS and the VABS domains for the nondemented groups. The results of these correlations are shown in Table 2.

The correlations in table 2 show that when scores from participants in all three groups are employed in the analysis there are significant correlations between both sub-scales of the A.A.D.S. and those of the D.M.R.. This demonstrates concurrent validity for the A.A.D.S.. Additionally, when scores from participants in all groups are analysed both the D.M.R. sub-scales and the A.A.D.S. sub-scales evidence strong correlations with domain scores for the V.A.B.S. and B.P.V.S. but not for age. This shows that both measures are not associated with age per se and the significant correlations with V.A.B.S. and B.P.V.S. for both measures are probably due to the influence of dementia. Significantly, inspection of the correlations for the A.A.D.S. and D.M.R. sub-scales with the V.A.B.S. domains when the Dementia Group is excluded reveals a different pattern for the two measures. While the D.M.R. evidences strong correlations with the V.A.B.S. domains in those who do not have dementia, similarly strong correlations are not evident for the A.A.D.S. sub-scales. This suggests that in
those who do not have dementia the A.A.D.S. sub-scales scores are less influenced by
degree of intellectual disability in comparison to the D.M.R. sub-scales.

To examine differences in the number, frequency, management and effect of
behavioural excesses and deficits between the groups, the mean total scores for each
of these subscales were compared. Table 3 shows the group means for these data and
the results of statistical comparisons.

The data presented in table 3 show significant differences between the groups on the
number and frequency of both deficits and excesses. Post hoc analyses reveal
significantly higher mean scores in the Dementia Group than both the Older and
Younger Comparison Groups on the four relevant subscales. A similar pattern is
evident for the scores on the Management of excesses and deficits subscales.
However, the Effect subscales differentiate less well between the groups. There are no
significant differences between the groups for Deficits whilst the analysis for
Excesses reveals a significantly higher score for the Dementia Group than the Older
Comparison Group but not the Younger Comparison Group. In summary, these data
show that behavioural excesses and deficits are associated with dementia, that the
management is more problematic and effect of deficits more pronounced in dementia
but the effect on the individual of these excesses and deficits is less clear

The final analysis focussed on the potential differences for Excesses and Deficits for
Management and Effect scores within and between the groups. As both the
Management and Effect scores are influenced by the number of Deficits and Excesses identified and the number of items in the subscales differs, the mean deficit and excess scores were derived for each participant to normalise the data for comparison. Mixed model analyses of variance tests were conducted to evaluate differences in management and effect scores between excesses and deficits and groups and potential interactions. The scores and results of the analysis are shown in table 4.

The data in table 4 show that for both management difficulty and effect there is no difference between excesses and deficits and no interaction with group. Thus, excesses and deficits appear to have a similar impact on carers and the individual regardless of age or dementia status. However, there is a main effect of group for both management difficulty and effect, with Scheffe post hoc analyses revealing a higher score for the Dementia Group in comparison to both the Older and Younger comparison groups. Thus, deficits and excesses associated with the presence of dementia give rise to greater management difficulty and effects on the individual. When this analysis was repeated using the frequency of deficits and excesses as covariates the main effect for management difficulty remained significant (F(2,31) = 4.71, p<.05) but there were no significant main effects or interaction for the effect on the individual. These analyses indicate that management difficulty for behavioural deficits and excesses is related to their presence but not their frequency but the effect on the individual is related to the frequency of deficits and excesses.
Discussion

In this study we have examined the presence and impact of behavioural deficits and excesses in adults who have Down syndrome and dementia using a design that has controlled for the potentially confounding effect of age. Neuropsychological measures previously employed in both cross-sectional and longitudinal studies of dementia in Down syndrome (Crayton, Oliver, Holland, Hall and Bradbury, 1998; Oliver, Crayton, Holland and Hall, 1998) demonstrated significantly poorer performance by the Dementia group than the group of comparable age, thus demonstrating integrity to allocation of participants to the Dementia group. Results of a widely used informant based measure (DMR, Evenhuis, Kengen and Eurlings, 1990) further supported group allocation.

The AADS measure developed for the study has robust inter-rater reliability for the six subscales. The evaluation of inter-informant reliability, as opposed to test-retest reliability, is important for an informant based measure that might be used prospectively when informants might change over time (see Oliver, 1999). Additionally, the subscales of the frequency of behavioural deficits and excesses both evidence good internal consistency and there is preliminary evidence for the concurrent validity of the frequency subscales for both deficits and excesses, as the total scores are significantly correlated with the most widely used informant based measure, the DMR (Evenhuis, Kengen and Eurlings, 1990). These analyses of the psychometric properties of the AADS are encouraging. However, the assessment of validity would be enhanced by prospective studies with supportive neuropsychological assessment and a larger sample is needed to generate normative data for age bands and degree of intellectual disability. Additionally, validity of the
constructs of management difficulty and effect on the individual warrant further
research. It is particularly important to further explore the effects on individuals of
behavioural deficits and excesses as the measure described here relied on the
perceptions of carers only. At present the AADS provides a useful appraisal of
specific behavioural deficits and excesses shown by adults with Down syndrome who
have dementia in terms of their frequency and their impact on carers and the
individual showing the behaviours

With regard to the validity of the AADS, of significance is the finding that the scores
on the frequency subscales for deficits is not correlated with scores on the Vineland
subscales when participants in the dementia group are excluded from the analysis,
although there is a significant negative correlation with the BPVS. This suggests that
performance on the deficit subscale of the AADS is not related to the overall level of
adaptive behaviour when dementia is not evident and, by implication, that the AADS
is sensitive to dementia related deficits. However, this conclusion warrants further
examination by comparing younger adults with Down syndrome and no dementia and
profound or severe intellectual disability with young and older groups with dementia
but a premorbid moderate or mild intellectual disability.

There is good evidence from administration of the AADS that both behavioural
deficits and excesses are demonstrated by adults with Down syndrome who have
developed dementia. This is in accordance with the case description literature and
reviews of behaviour change and difference (Prasher and Filer, 1995; Dalton and
Wisniewski, 1990; Newroth and Newroth, 1980; Oliver, Crayton, Holland and Hall,
2000; Adams et al., 2008). The measure might usefully be employed as an adjunct to
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neuropsychological tests and routine examinations for the diagnosis of dementia, preferably as part of a battery of repeated assessments. However, the measure cannot employed as a diagnostic instrument. The issues of baseline assessment, monitored change and routine examination are critical to differential diagnosis.

The examination of the impact of behavioural deficits and excesses on the individual and carers reveals greater management difficulty and effect on the individual for the dementia group. These results confirm previous research that has demonstrated an association between behavioural change associated with dementia and the effect on carers and the individual’s life experiences (Oliver, Crayton, Holland and Hall, 2000). Secondary analyses of these data show that when controlling statistically for the frequency of deficits and excesses, there is no difference between deficits and excesses in terms of management difficulty for carers or the effect on the individual. Additionally, the management difficulty of deficits and excesses for carers is not related to the frequency of the deficits and excesses but this is not the case for the effect on the individual. In combination, these analyses suggest that the behavioural deficits and excesses associated with dementia in persons with Down syndrome are similar with regard to both degree of management difficulty for carers and the effect on the individual and that the management of deficits and excesses is related to presence as opposed to frequency.

There are a number of areas that might be pursued to extend research into behavioural change and difference in dementia. The AADS might usefully supplement neuropsychological assessment employed in research into dementia, particularly in prospective studies and when floor effects become evident in testing. Prospective
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studies might elucidate the pattern of specific deficits and excesses that emerge with the progression of dementia. However, this will depend on item reliability being established for the AADS. A larger scale study would allow factor analytic approaches to be adopted and subsequently for the analysis of change over time to be examined on empirically derived subscales. Finally, the subscales that appraise management difficulty and the effects on the individual of the presence and frequency of deficits and excesses might usefully be employed to inform service delivery and also be used to evaluate service design, carer training and psychosocial interventions.
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**References**


Dunn, Dunn, Whetton & Pintilie (1992). *The British Picture Vocabulary Scales*


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<table>
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<tr>
<th></th>
<th>Younger comparison group (A)</th>
<th>Older comparison group (B)</th>
<th>Dementia group (C)</th>
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<th>Posthoc analysis</th>
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Table 1. Mean scores, standard deviations and results of statistical comparisons for groups on neuropsychological and informant based measures of dementia.
Table 2. Pearson correlation coefficients for participants in all groups and the non dementia groups only for the AADS and DMR subscales with the VABS subscale and BPVS scores and age. (* = p. <.05; ** = p. <.01)

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Table 3. Mean scores and standard deviations of the number and frequency of behavioural deficits and excesses and the management difficulty and effects on the individual of behavioural deficits and excesses broken down by group with results of oneway analyses of variance comparisons and posthoc analyses.

<table>
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<tr>
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<th>Younger comparison group (A)</th>
<th>Older comparison group (B)</th>
<th>Dementia group (C)</th>
<th>F(2, 33)</th>
<th>Posthoc analysis</th>
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<td>3.48</td>
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<td>Effect (total)</td>
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<td>Excesses</td>
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<td>0.62</td>
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<td>1.59</td>
<td>1.50</td>
<td>3.12</td>
<td>7.92</td>
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Table 4. Mean scores, standard deviations and results of the mixed model ANOVA analysis for the groups on the management difficulty and effects on the individual subscales of the AADS.