Multidimensional apathy in ALS: Validation of the Dimensional Apathy Scale

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ABSTRACT

Apathy is a prominent symptom of Amyotrophic lateral sclerosis (ALS), but measurement is confounded by physical disability. Furthermore it has been traditionally measured as a unidimensional symptom despite research demonstrating a multifaceted construct. The new Dimensional Apathy Scale (DAS) has been specifically designed for patients with motor disability to measure three neurologically based subtypes of apathy; Executive, Emotional and Initiation. We aimed to explore this behavioural symptom by examining the substructure of apathy in ALS and to determine the reliability and validity of the DAS in patients and their carers.

Method: Patients and carers were recruited through the national Scottish Motor Neurone Disease Register and were asked to complete the DAS, the standardised Apathy Evaluation Scale, and the Geriatric Depression Scale- Short form. 83 ALS patients, 75 carers and 83 sex-age-education matched controls participated.

Results: When compared to healthy controls, patients showed a significant increase in apathy on the Initiation subscale, and were significantly less apathetic on the Emotional subscale. Scores on the DAS patient and carer versions did not significantly differ. Internal consistency reliability, convergent and discriminant validity were found to be good for the DAS subscales. There was no association between the DAS and functional disability using the ALS Functional Rating Scale.

Conclusion: Apathy in ALS is characterised by a specific profile of increased Initiation apathy and reduced Emotional apathy. The DAS is a reliable and valid measure for the assessment of multidimensional apathy in ALS.

INTRODUCTION

Apathy is defined by decreased motivation towards goal directed behaviours,[1] and occurs as a symptom of a variety of different psychiatric and neurodegenerative diseases,[2, 3]. Studies have shown that apathy occurs in 30% to 60% of ALS patients and is the most prominent behavioural symptom of the disease,[4-7]. This behavioural change in ALS has most commonly been detected by the Frontal Systems Behavior Scale (FrSBe) apathy subscale,[8]. However, this tool is not specifically designed to assess patient populations with physical disability and may exaggerate behavioural symptoms as responses to some items are reliant on effective motor functions,[9]. Other tools which have been used to detect behavioural change in ALS are the Cambridge Behaviour Inventory- Revised,[10], the ALS- Frontotemporal Dementia- Questionnaire,[11] and, finally, the Motor Neurone Disease Behavioural Instrument,[12]. These measure apathy as a part of just one of many behavioural or psychiatric disturbances and lack a detailed analysis of symptoms.

Marin originally defined apathy as a multidimensional concept that is composed of factors relating to cognitive, behavioural and emotional domains,[13, 14]. Although no previous scales have been comprehensively designed to directly measure the subdomains of apathy, factorial analysis of current scales has revealed a sub structure similar to that of Marin's original conceptualisation in Parkinson's disease,[15], observing a traditional triadic substructure. Additionally, in a comparison of Alzheimer's disease and Frontotemporal dementia, apathy was found to have differing profile characteristics,[16]. Specifically, the Neuropsychiatric Inventory – apathy subscale items showed that

behavioural variant Frontotemporal dementia patients were reported as more frequently showing lack of initiation, decreased emotional output and diminished interest towards friends or family, when compared to Alzheimer's disease patients. However, no such research has been undertaken in ALS as yet.

Traditional instruments measure apathy as a unidimensional symptom despite clear evidence of a multidimensional substructure. The new Dimensional Apathy Scale (DAS),[17] was designed in a cohort of healthy adults for use in neurodegenerative disease patient populations with motor symptoms. The DAS was specifically designed to measure three neurobehavioral apathy subtypes,[18, 19] through theory based analysis and selection of items. It assesses apathetic impairments associated with planning, attention or organization (Executive), emotion integration (Emotional) and self-generation of behaviours or cognition (Initiation). The DAS has been shown to have good internal consistency reliability, with subscales having a moderate relationship to depression and is the only method that comprehensively measures apathy subtypes.

Apathy and depression have been reported to have a variable relationship, with some studies describing them as distinct factors and others reporting an association.[20, 21]. Lack of interest, insight and reduced energy have been suggested as overlapping characteristics of apathy and depression, with dysphoria, suicidal ideation and helplessness being depression specific and indifference, diminished initiation and poor persistence being apathy specific,[22-25]. Documenting the relationship between apathy and depression

in neurodegenerative disease remains important as they may be differentially affected or overlapping symptoms misattributed.

The aim of this study was to determine why apathy is a particularly prominent feature of ALS through an exploration of the substructure of this behavioural symptom. In addition we aimed to determine the psychometric properties of the DAS, namely the validity as assessed against a standardised measure of apathy and its association with depression and disease related disability.

METHOD

Procedure and Participants

El Escorial criteria diagnosed ALS patients and their carers were recruited from the national Scottish Motor Neurone Disease Register. Patients were recruited via postal survey. Patients with Primary lateral sclerosis or Progressive muscular atrophy were not included in the study. Carers were recruited via the patients through a chain-referral sampling method. Patients were anonymously pre-screened for severe disability as a result of disease progression that would hinder completion of the survey, pre-existing dementia, severe diabetes, epilepsy, alcohol/substance- related disorders, severe head injury (that required intensive care setting hospitalisation), traumatic brain injury (inclusive of subarachnoid haemorrhage) and any other significant medical illness (such as stroke).

Of 190 ALS patients who were contacted, 46.8% patients and 44.2% carers returned the postal survey. Of the participants who returned the survey, 3.5% of

patients and 5.9% of carers returned incomplete questionnaires and were subsequently excluded. A further 3.5% of carers did not have a matched patient completed form and were also excluded. This resulted in 83 ALS patients and 75 carers being included in the study.

400 healthy control participants completed the study. They were recruited via web-based survey and were mostly from the University of Edinburgh Psychology Departmental Volunteer Panel. Before recruitment, they were additionally pre-screened for any serious physical or mental health issues. 83 healthy controls were subsampled from this pool to match the patient group for sex distribution, age and years of education.

Ethical approval was obtained from the National Health Service (NHS) South East Scotland Research Ethics Committee 02 and the School Philosophy, Psychology and Language Sciences (PPLS) Ethical Committee.

Apathy

The DAS,[17] is multidimensional scale composed of 24 items constituting 3 subscales assessing Executive, Emotive and Initiation apathy. Items were scored using a 4-point Likert scale based on the frequency of occurrence in the last month. The minimum score for each subscale is 0 (least apathy) and the maximum 24 (most apathy), with a total score of 72. It was shown to have good internal consistency reliability and to have a weak to moderate relationship with depression. A self version has been previously reported by the authors and a

carer version was adapted specifically for this study[1]. Normative data was subsequently used to suggest abnormality level cut-offs for each subscale based on ≥2 SD above the mean (see Table 1).

Table 1. Normative Data on DAS (N=83)

	Mean (SD)	Range	Abnormality cut-off
DAS Executive subscale	5.9 (4.2)	0–17	14
DAS Emotional subscale	8.8 (2.9)	3–19	15
DAS Initiation subscale	9.5 (3.5)	1–17	16
DAS Total	24.1 (7.3)	8-42	39

DAS=Dimensional Apathy Scale, the maximum for each subscale is 24 and the total is 72

The Apathy Evaluation Scale (AES),[14] is composed of 18 items and is scored on a 4 point Likert scale based on frequency of occurrence in the last month. It produces one composite score, where the minimum score is 18 (least apathy) and maximum score of 72 (most apathy). The recommended cut-offs for the self version is based on 2 standard deviations above the mean,[14]. For our control sample this was 39, which is consistent with the original study. The carer version cut-off was defined as 40. The AES is a well-established method of detecting apathy, and has been shown to be both valid and reliable,[3, 25]. Both a carer and patient versions were available.

Depression

Geriatric Depression Scale-Short form (GDS-15),[26, 27] is an abridged version of the Geriatric Depression Scale. It is composed of 15 items, with statements

[1] See Supplementary material

graded on a dichotomous, yes-no scale within the previous week. The minimum score is 0 (not depressed) with the maximum being 15 (most depressed). Recommended cut-offs for the patient self rated version are disputed but highest consensus in the literature is a cut-off of >6 for presence of depressive symptoms,[28]. Both a patient and carer version were used.

Disease Related Disability

The ALS Functional Rating Scale-Revised (ALSFRS-R),[29] is a 12 domain global functioning and disability measure specifically designed for assessment of ALS patients. Each domain scored on a 5 point scale with the total score ranges from 0 (maximum disability) and 48 (normal motor function).

Statistical Analysis

R and SPSS statistics 19.0 was used to analyse the results. Independent t-tests and Chi-square tests were used to compare demographics, symptom frequency and clinical variables between ALS patients, and controls, DAS Subscale impaired patients and unimpaired patients. Internal consistency reliability was assessed using Cronbach's Standardized alpha. Validity was examined using correlational analysis (Holm corrected Pearson's r). A 2 x 3 Mixed design Analysis of variance (ANOVA) was used for comparison of Group (Patients vs Carers/Controls) and DAS subscale (Emotional vs Executive vs Initiation). Post hoc Independent t-tests were used for subscale and factor comparison.

RESULTS

Table 2. Demographic and descriptive data for ALS patients (N=83), their carers (N=75) and controls (N=83)

	ALS Patient	ALS Carer	Control	Patient vs Control *p*-value
DAS Total Score (mean, SD)	25.7 (10.6)	27.3 (12.8)	24.1 (7.3)	n.s.
DAS Total (Apathy/No Apathy)	12/71	14/61	2/81	<.01
AES Score (mean, SD)	30.9 (8.6)	32.8 (10.9)	28.9 (5.0)	n.s.
AES (Apathy/No Apathy)	20/63	18/57	1/82	<.001
GDS-15 Score (mean, SD)	5.9 (4.2)	6.7 (4.7)	2.5 (2.8)	<.001
Age (mean, SD)	64.6 (10.5)		63.7 (13.0)	n.s.
Years of Education (mean, SD)	13.5 (3.4)		14.4 (2.7)	n.s.
ALSFRS-R Score (mean, SD) †	37.7 (6.2)			
Age of onset (mean, SD) years†	59.6 (11.0)			
Disease duration (Median, IQR) months††	66.5 (71)			

ALS=Amyotrophic lateral sclerosis; SD=standard deviation; DAS=Dimensional Apathy Scale; AES=Apathy Evaluation Scale; GDS-15=Geriatric depression scale- Short form; ALSFRS-R=ALS Functional Rating Scale-Revised; n.s.=not significant; IQR=Interquartile range
† N=32
†† N=62

Background Information

There was no significant difference between patients and controls on age and years of education (see Table 2). Sex distributions were matched, with 57 males and 26 female participants in both samples. The most common carer relationship to the ALS patient was a spouse.

Apathy (AES) and Depression

The results of the AES revealed that the total score was not found to significantly differ between patients and controls, however the number of individuals who fell above the suggested cut-off for abnormality did, with significantly increased levels of abnormal apathy in the patient group (see Table 2).

In this sample, according to the AES, 24% of patients self rated and 24% of carers rated patients as apathetic. Depression rates were higher, with patient-rated depression at 39% and carer-rated depression at 44%. Additionally, patient reported depression scores were found to be significantly different when compared to controls and just under borderline diagnosis level (see Table 2). Furthermore the AES was found to be positively correlated with GDS-15 in patients ($r(73) = .65$, $p < .001$) and carers ($r(81) = .67$, $p < .001$).

Patient (self-rated) and Carer-rated comparison on the DAS

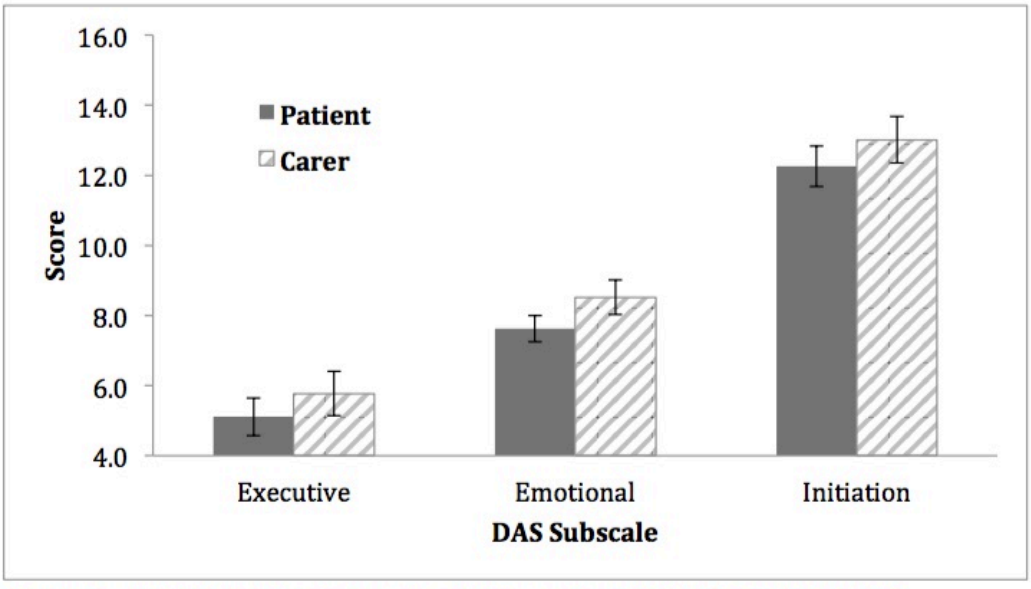

Figure 1. ALS self rated and carer rated scores on DAS Subscales (N=75)

The comparison between Group (Self vs Carer) and DAS subscales is presented in Figure 1. There was no significant interaction between Group and the DAS Subscales with carer's ratings being only, on average 0.7, points higher than patient self ratings. However, there was a significant main effect relating to DAS subscales ($F(2,296) = 160.30$, $p < .001$). Post hoc independent t-tests within groups showed that in patients the DAS Executive subscale differed significantly from Emotional subscale ($t(164) = 3.53$, $p < .01$) and the Initiation subscales ($t(164) = 9.11$, $p < .001$), in addition there was a significant difference between Emotional and Initiation subscales ($t(164) = 7.19$, $p < .001$). In carers, a similar relationship between subscales was found where the DAS Executive and Emotional subscales differed significantly ($t(148) = 3.42$, $p < .01$), along with Executive and Initiation subscales ($t(148) = 7.93$, $p < .001$) and Emotional and Initiation subscales ($t(148) = 5.45$, $p < .001$). There was no significant difference on the DAS total score between patient and carer ratings.

Patient (self-rated) and Healthy Control comparison on the DAS

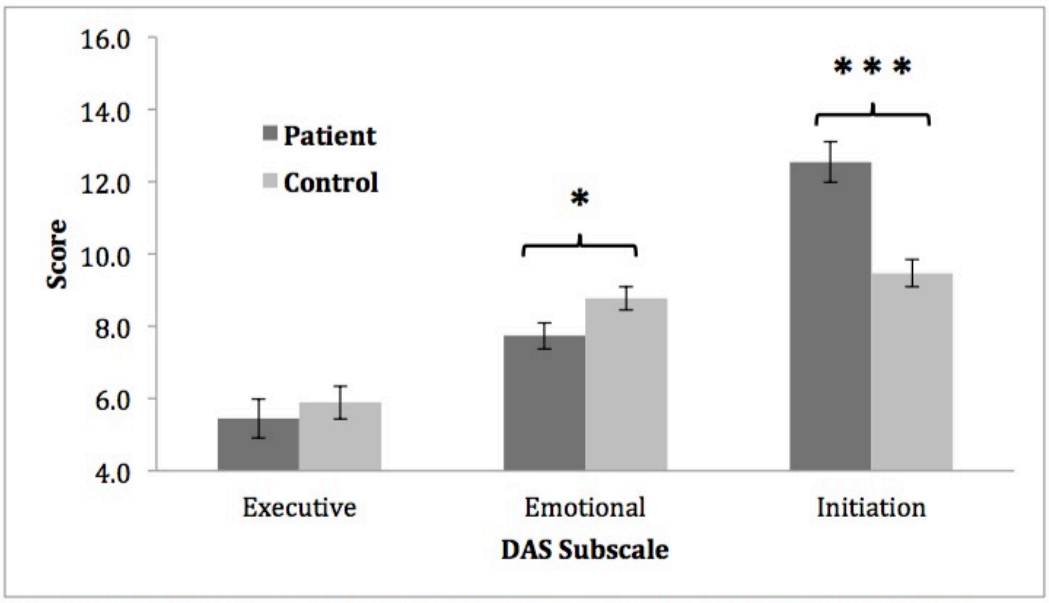

Figure 2. ALS self rated and healthy control scores on DAS Subscales (N=83)
***** *p*< .001, * *p*< .05**

A comparison of group (Self vs Control) and DAS Subscale, showed a significant main effect for DAS subscales ($F(2,328) = 107.16$, $p < .001$) indicating dissociations between subscales (see Figure 2). In addition, a significant interaction was also found between Group and DAS subscales, reflecting differential performance between patients and controls on the different subscales ($F(2,328) = 18.56$, $p < .001$).

Further post hoc t-tests showed that Initiation was the only subscale in which patients were significantly more apathetic than controls ($t(164) = 4.52$, $p < .001$). Additionally, on the emotional apathy subscale, patients scored significantly lower when compared to control participants ($t(164) = 2.15$, $p < .05$). The DAS total score did not significantly differ between patients and controls.

Psychometrics

The overall DAS Cronbach's Standardized alpha values were 0.86 for the self-version and 0.90 for carer-version, which can be interpreted as good and excellent [3], respectively. There were good internal consistencies for both the Executive (self = 0.86, carer = 0.88) and Initiation (self = 0.83, carer = 0.86) subscales, with the poorest being for self DAS Emotional subscale at 0.43. However, the carer Emotional subscale showed a higher internal consistency at 0.65.

Table 3. Patient (self rated) and carer rated DAS subscale correlations compared to standardised, self-report apathy (AES) and depression (GDS15) measures

Self (N=83)	AES	GDS-15
DAS Executive subscale	0.76***	0.61***
DAS Emotional subscale	0.21	0.20
DAS Initiation subscale	0.79***	0.61***
DAS Total	0.80***	0.67***
Carer (N=75)	AES	GDS-15
DAS Executive subscale	0.82***	0.64***
DAS Emotional subscale	0.44***	0.24*
DAS Initiation subscale	0.79***	0.65***
DAS Total	0.85***	0.64***

DAS=Dimensional Apathy Scale; AES=Apathy Evaluation Scale; GDS-15=Geriatric Depression Scale- Short Form ***p*<.001*****, *p*<.05***

The DAS Subscales positively correlated with the AES, with moderate correlations with the Emotional subscale and strong correlations with the Executive and Initiation subscales (see Table 3). Similarly the carer DAS subscales were more positively correlated with the AES compared to the GDS-15.

Disease related disability

A total of 32 patient's and, of those, 27 carer's ALSFRS-R scores were acquired. The ALSFRS-R did not significantly correlate with the self DAS Executive subscale, Emotional subscale, Initiation subscale and the total score. When compared to the carer version of the DAS, the ALSFRS-R was also not significantly correlated with any of the subscales and the total score.

Diagnostic cut-off for the DAS

Using the abnormality level cut-offs in Table 1, 28% of ALS patients were impaired on at least one apathy subscale, of which 61% were impaired on the Initiation subscale only and 39% were impaired on Initiation and one other subscale (30% Executive and 9% Emotional). In carers, 43% were impaired in at least one subscale, where 56% were impaired on the Initiation subscale only, 10% on the Executive subscale only and 6% on the Emotional subscale only. A further 16% displayed Initiation and Executive apathy with 6% showing Initiation and Emotional apathy. A total of 6% showed apathy on all 3 subscales.

Table 4. Comparison of patients (carer-rated) impaired on ≥1 DAS Subscales (N=25) to patients unimpaired on all subscales (N=34)

	≥1 Subscale Impairment	Unimpaired	*p*-value
ALSFRS-R (Mean, SD)	38.8 (4.2) †	36.6 (8.8) ††	n.s.
Disease duration (Median, IQR)	45 (53)	98 (74)	<.05
Age of Onset (Mean, SD)	57.2 (11.4)	60.6 (10.6)	n.s.
Site of Onset (%)			
Bulbar (N=15)	26.7%	73.3%	
Lower Limb (N=21)	47.6%	52.4%	

Upper Limb (N=19)	47.4%	52.6%	
Mixed (N=4)	50%	50%	

ALSFRS-R=ALS Functional Rating Scale- Revised; SD=standard deviation; n.s.=not significant; IQR=Interquartile range

† **N=10**

†† **N=11**

Table 4. is a comparison clinical variables in apathetic and non-apathetic patients as defined by impairment on at least one DAS subscale as rated by the carer. Of the 75 carer rated DAS scores, 16 patient's clinical variables were unavailable, resulting in a total of 59 patient's clinical variables being used. There was a significant difference between the two groups on disease duration, wherein ≥1 Subscale Impairment patients had the disease for a shorter time at assessment. The proportion of impaired and unimpaired patients with lower limb, upper limb and mixed onset was relatively equal, although those with bulbar onset were more likely to be unimpaired than impaired. However, frequency distributions did not differ using Chi-square statistics (which excluded the mixed onset in the analysis).

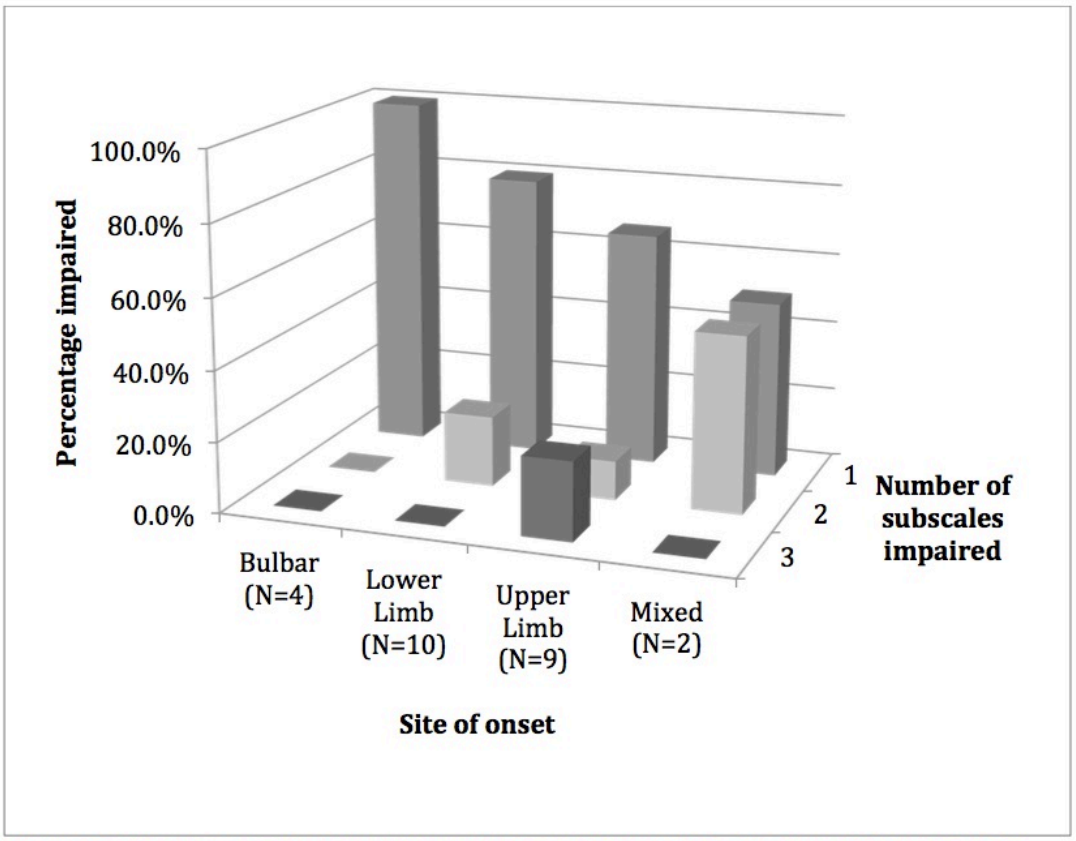

Figure 3. Frequency of patients impaired on different of numbers DAS subscales (carer-rated), divided by site of onset (N=25)

Figure 3 shows a further subdivision of impairment (impaired on all three subscales, impaired on two subscale and impaired on one subscale) based on site of onset. All bulbar onset patients who were impaired on the DAS showed impairment on one subscale, while those with lower limb onset and upper limb onset were impaired on 2 or more subscales. The only 2 instances of global impairment were in the upper limb onset group. The most common impairment on one subscale across all groups was on the Initiation subscale.

DISCUSSION

The DAS was shown to be sensitive to apathy in ALS, where 28% of patients and 43% of carers in our sample showed abnormal levels of apathy on at least one

subscale. The prevalence of apathy reported here is slightly lower than previous studies, where apathy was reported between 30-60% of the patients sampled [4-7]. However previous studies have utilised measures, which have not been designed for physical disability and where symptoms of apathy may be exaggerated by motor dysfunction. The more conservative estimate reported here might be therefore a more accurate reflection given that the DAS was designed to measure apathy independent of physical disability.

The most prevalent DAS subscale impairment was in Initiation with ALS patients showing significantly increased Initiation apathy compared to controls. Initiation apathy consists of a lack of self generated behaviour and cognition,[22]. Such processes are also dependent on intact executive functions, which are known to be affected in ALS,[30].

Stuss proposed a model of executive functions that includes the concept of energization, defined as diminished initiation and sustainment of responses to tasks,[31, 32] and which clearly has overlap with Initiation apathy. Energization deficits are often observed as decreased output during verbal fluency tasks, increased errors and slow response time for Stroop tasks. This verbal fluency deficit is a characteristic feature of the cognitive profile in ALS,[30] and has been found to correlate with apathy as measured by the FrSBe,[4]. Further research using the Edinburgh Cognitive and Behavioural ALS Screen (ECAS),[33] found a high prevalence of the fluency deficit in ALS patients and, additionally, reported apathy as the most prevalent behavioural impairment although the behaviour screen contains just one item on apathy.

Initiation apathy is akin to one of Levy and Dubois apathy subtypes- Auto Activation,[18]. This type of apathy has been related to lesions in the medial prefrontal region, anterior cingulate cortex and caudate nucleus. Brain imaging studies have found abnormalities in ALS patients in the anterior cingulate cortex to be related to apathy,[34] and the caudate nucleus has also been shown to be affected in ALS, and it was posited that this may be involved with the mediation of motivation,[35]. Similarly verbal fluency deficits (the key feature of the energization deficit) has been related to dysfunction of similar prefrontal regions,[36, 37].

It is of note that the Executive subscale showed no difference between patients and controls. This subscale assesses apathy as a result of poor planning, organisation and attention. At first glance this appears inconsistent with research demonstrating executive dysfunction is a common feature in non-demented ALS patients as revealed through neuropsychological evaluation. However there are very few reports of this deficit manifesting behaviourally or having an impact on daily functioning in non-demented ALS patients,[38, 39]. Current research within ALS distinguishes between the types of executive functions, which are impaired with distinct neural substrates,[37]. The distinction between Initiation and Executive apathy is further supported by Levy and Dubois,[18, 19], and our own data driven approach, which also separated these two factors,[17] in healthy controls. The findings here demonstrate that the processes of initiation of thought and action appear to be the crucial element, which underlie the behavioural manifestation of apathy in ALS.

The finding of lower Emotional apathy in ALS, when compared to controls, may be related to dysfunction in emotional processing, theory of mind and social cognition in ALS which has been recently documented,[9]. A deficit in social cognition has been associated with apathy, although cognitive impairment was associated with an increased level of apathy,[7]. Future studies may demonstrate whether this social cognitive impairment is related to increased Initiation apathy specifically. Other factors must be considered in relation to this lack of emotional indifference in ALS patients. These may include increased sensitivity to emotion associated with reaction and impact of having a terminal disease. However it should be noted that there was only a weak correlation between emotional apathy and depression, which was in a positive direction, i.e. the more apathetic the more depressive symptoms. Conversely the patients with lower emotional apathy tended towards fewer depressive symptoms. However ALS patients do show a higher incidence of emotional lability, which is involuntary occurrence of positive (laughing) and negative (crying) emotions,[40], which may be regarded as the antithesis of Emotional apathy. Emotional lability was not recorded here and it may serve as an underlying factor in the reduced emotional apathy in ALS patients.

There were some differences in clinical variables between patients with apathy and those without when broken down by the impairment on the DAS. Patients with apathy as defined by impairment on one or more subscales had the disease for a shorter number of months when compared to unimpaired patients. This is consistent with the finding that apathy is the most common behavioural change

associated with the onset of disease, as measured by the FrSBe,[4]. It would also be of interest to explore how apathy develops through progression of the disease. Cognitive impairment has been shown to be a negative prognostic factor,[41] and the current research poses the question of whether this is also true of patients with apathy. Prospective studies may also determine the course of this symptom and whether apathy characterises a distinct subgroup of patients.

Further break down of impairment showed that all patients irrespective of site of onset were most commonly impaired on one subscale, that being Initiation. This provides further evidence that the Initiation deficit is that which defines apathy in ALS. It is also of interest that the patients with bulbar onset did not show a greater vulnerability to apathy, and therefore does not support previous associations of cognitive and behaviour change and bulbar symptomology,[42]. Given that functional disability data was only available on a subgroup of patients, although no correlations emerged here, the relationship between this behavioural symptom and physical dysfunction and progression should be further explored.

The DAS was found to be a psychometrically robust instrument for detecting apathy in ALS, with very little difference between patient and carer ratings. There was a consistent dissociation between the DAS subscales in patients, carers and also controls, providing evidence for the DAS measuring distinct subtypes of apathy. Previous reviews of assessment methods in ALS outlined one of the main issues of behavioural measurement is that it is confounded by motor

dysfunction,[9]. The DAS was specifically designed to account for these motor symptoms and the current findings of apathy were independent of functional disability supporting its usage in patients with motor symptoms. Furthermore, our control samples apathy levels, as measured by the AES, were comparable to that of Marin's in his original validation of the AES (Mean=28.1, Standard Deviation=6.4),[14] making our control group no more apathetic than in Marin's original study and therefore suitable for comparison to the patients group.

The internal consistency reliability for the whole DAS was good, with it being higher for the carer (0.90) than the self version (0.86). When examining subscale internal consistency reliability, we found that the self and carer Executive and Initiation subscales were found to be very reliable however, the self DAS Emotional subscale was found to be poor. Patients' self awareness may be affecting performance on the self rated questionnaire, and there is some evidence of poor insight in ALS patients,[43]. However it is at present unclear why this would differentially affect the emotional component and this interaction may be an area for future research. The internal consistency for the carer Emotional subscale was markedly higher, suggesting that the carer assessment might be a more informative method of assessing Emotional apathy.

Our study found that associations with the DAS apathy subscales and the standardized apathy measure (AES) were on average more positive and stronger than with the depression measure (GDS-15), resulting in a good convergent and discriminant validity of the DAS. When looking specifically at the Emotional apathy subscale, the self version was marginally more positively associated with

the AES than the GDS-15, however the difference was minor. The carer DAS Emotional subscale more positively and strongly associated with the AES than the GDS-15, showing in a good validity and reliability of this subscale. The prevalence of depression in our patient sample was found to be in line with that of other studies,[44].

A caveat of all apathy research is volunteerism, where participants who participate in studies are likely to be more motivated and less apathetic. Our study may, therefore, be underrepresenting the prevalence of apathy in ALS. However the response rate of the current research (46.8% patients and 44.2% carers) is as good if not better than other MND studies,[6].

To conclude, the DAS is a psychometrically valid and reliable instrument for detecting dissociable apathy subtypes, independent of physical disability. Apathy in ALS seems to be defined by specific impairments in initiation of cognition and behaviour. Additionally, patients seem to exhibit a lack of Emotional apathy. These novel findings suggest that apathy in ALS has a specific profile relating to initiation and emotion. The relationship between these subtypes and cognitive, behavioural and emotional change should be further explored. Furthermore the DAS is appropriate to use to determine different apathetic profile impairments in other neurodegenerative diseases, in which apathy is most prevalent. Future research should look to investigate the neuropsychological correlates associated with different apathetic subscale profiles in addition to quality of life and both patient and caregiver burden with the aim of directing care.

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COMPETING INTERESTS

None.

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