

Introduction

Amyotrophic lateral sclerosis (ALS) affects both motor and cognitive functions. Cognitive impairments include impaired executive function, attention, and verbal fluency [1-2]. Deficits in confrontation naming also have been reported [3]. However, despite the evidence of cognitive decline in areas that correlate with semantic deficits in both confrontation and generative naming paradigms, few studies have focused specifically on language function per se in ALS. Strong et al. [3] in a prospective study of participants with ALS performed a battery of language assessment items embedded in a comprehensive protocol of motor and neuropsychological measures. They reported early manifestations of cognitive impairment in ALS including confrontation naming and word generation. They also noted a significant occurrence of verbal and semantic paraphasia error types. Collectively, these studies indicate semantic access is impaired in ALS. The current study attempts to extend these findings by evaluating language production in a discourse paradigm using two elicitation tasks that vary by degree of semantic constraint and outcomes indicating word-finding problems

Method

Participants

All participants received a clinical diagnosis of ALS (El Escorial criteria) [4] and were selected such that disease duration was less than one year from time of clinical symptom onset. Participants were recruited as a convenience sample by the director of the Motor Neuron Diseases clinic at the London Health Sciences Center, University Campus, London Canada. There were sixteen ALS and twelve control participants. Controls included spouses or relatives of the participants with ALS.

There were fourteen participants with classical sporadic ALS and two with familial

ALS. Five presented with bulbar signs and symptoms at disease onset. Eleven presented with either upper limb (n=6) or lower limb (n=3) signs and symptoms at disease onset. There were 10 men and 6 women whose age at onset ranged from 34 to 68 years ($M=52.8 \pm 9.15$). They were all native speakers of English and their education ranged from 10 to 22 years ($M= 14.9 \pm 3.22$).

There were seven men and five women control participants. None had a history of neurological disease, including ALS, psychiatric illness or other medical or neurological conditions. All controls were native speakers of English whose ages ranged from 34 to 63 years ($M=53.3 \pm 8.03$). Their education ranged from 10 to 19 years ($M=13.4 \pm 2.60$).

Procedure

Discourse samples were obtained from each ALS and control participant in a face-to-face topic-directed interview (TDI) and using the Cookie Theft Picture description task from the *Boston Diagnostic Examination* [5]. The TDIs were initiated by the examiner using the starter phrase “Tell me about _____” and included the following five topics: 1) your family, 2) where you were born and raised, 3) your health right now, 4) the jobs you had or the work you did, and 5) what do you do each day. Participants were shown the Cookie Theft picture and prompted with the open-ended request: “Tell me everything that is going on in this picture”. The outcome measures included percent correct information units (CIU) [6] and seven indices reflecting word finding problems [7].

Inter-rater percent agreement for total number of words transcribed ranged from 80.2% to 100% ($M = 96.16$, $SD = 6.57$) while percent agreement for utterance segmentation ranged from 83.3% to 100% ($M = 92.35$, $SD = 5.24$). Inter-rater reliabilities for speech intelligibility and rate were strong for both intelligibility (.875) and rate (.972).

Correlational analysis indicated no significant correlation between intelligibility or rate and the total number of verbal fluency items generated for participants with ALS and controls in a parallel study with these participants thus minimizing the effects of rate and intelligibility on CIU and naming error variables. Alpha level was set at 0.05 while values between 0.05 and 0.10 were regarded as approaching significance.

Results

The mean total words produced, mean total CIU, and standard deviations for the ALS and the control participants appear in Table 1. The mean percentage CIUs for ALS participants for the Cookie Theft sample is 59.39% ($SD \pm 19\%$) and for the TDI sample is 89.01% ($SD \pm 5\%$). The mean percentage CIUs for control participants for the Cookie Theft sample is 79.23% ($SD \pm 21\%$) and for the TDI sample is 92.10% ($SD \pm 4\%$).

The mean percentage CIUs for all participants and tasks was analysed using a mixed ANOVA with between-participants/within-participants variables for group (ALS vs. Controls) and discourse task (Cookie Theft vs. TDI). There was an interaction of group x discourse task $F(1,25) = 5.545, p = < .027$. There was a main effect of group $F(1,25) = 6.969, p = < .014$ with the ALS group producing lower percent CIUs than controls.

The mean percentage instances of word retrieval problems by instance type for the ALS and control groups for both the Cookie Theft and the TDI samples appear in Table 2. To analyze the effect of group (ALS vs. Control) on the percent of instances of word retrieval problems, the mean percent for each type of instance of word retrieval problem separately for the Cookie Theft and TDI data were subjected to a multivariate ANOVA. There was a main effect of group for the Cookie Theft $F = 4.931, p = < .003$. The F and p values for each instance type for the Cookie theft data appear in Table 3.

There was a main effect of group for the TDI data $F = 2.171$, $p = <.090$. The F and p values for each instance type for the TDI data appear in Table 4.

Discussion

With a conservative threshold set at 60% CIUs, 8/15 ALS participants in the current study fall below this threshold on the Cookie Theft sample compared to 3/12 controls. The percentage of CIUs was significantly higher among participants with ALS during the TDI discourse task vs. the Cookie Theft task suggesting that the nature of the discourse task interacts with language efficiency for individuals with ALS.

Discourse elicitation tasks using pictures such as the Cookie Theft stimulus in this study arguably are more semantically constrained placing increased demands on lexical access. As such, the findings of this study may suggest that the reduced efficiency of language output in ALS reflects semantic deficits. The indices of word finding problems analysis from this study supports this suggestion with significant differences in instance type reported for word reformations (revising utterances), empty utterances, substitutions (semantic paraphasias or substituting non-specific words “stuff”), and delays.

The TDI discourse task is less semantically constrained. Participants are required to speak only within a defined topic but are relatively unrestricted in their word choices. The significantly higher CIUs produced during the TDI task for the ALS group may reflect the lower and less constrained semantic demands of this task. The instance type for word finding problems also was different with the only instance type reaching statistical significance being insertions (comments on the language process). Our findings provide preliminary evidence that individuals with ALS without dementia have deficits in language efficiency suggestive of semantic access difficulties.

References

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Table 1 Mean total words and total CIUs

Measure	Total words	SD	Total CIU's	SD
CT-ALS	89.7	41.6	51.5	30.0
CT-Control	80.7	58.4	56.2	38.1
TDI-ALS	932.4	480.2	810.1	389.2
TDI-Control	929.3	580.4	845.1	513.9

Table 2 Mean percentage instances of word retrieval problems

Instance Type	ALS-Cookie Theft	ALS-TDI	Control-Cookie Theft	Control-TDI
Repetition	9.4	35.8	5.3	38.2
Word reformation	13.7	32.8	1.8	33.9
Empty words	11.6	6.0	1.0	4.3
Insertions	13.0	8.3	16.0	1.8
Substitutions	3.4	14.1	1.0	14.8
Delays	3.1	1.0	0	3.7
Time Fillers	1.5	2.6	1.0	3.3

Table 3. *F* and *p* values for Instances of word finding problems for Cookie Theft data

Instance type	F value	p values
Repetition	.699	.411
Word reformations	9.025	.006
Empty utterances	7.155	.013
Insertions	.231	.635
Substitutions	3.400	.077
Delays (> than 5 sec.)	4.865	.037
Time fillers	.407	.530

Table 4. *F* and *p* values for Instances of word finding problems for TDI data

Instance type	F value	p values
Repetition	.389	<.539
Word reformations	1.199	<.284
Empty utterances	.000	<.986
Insertions	5.190	<.032
Substitutions	.034	<.856
Delays (> than 5 sec.)	1.915	<.179
Time fillers	.129	<.723

Table 4.