



The Dissociation of Dysarthria from Cognitive Dysfunction in ALS



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INTRODUCTION

Recent studies in ALS suggest structural and pathologic changes extend beyond motor neurons and correlate with cognitive dysfunction. The prevalence and types of cognitive impairment reported in association with ALS have varied greatly. Current studies have described some degree of cognitive impairment in 55 to 75% of patients with ALS. The majority of studies have focused on the overlap between ALS and frontotemporal dementia (FTD), although cases of less severe cognitive impairments have been described. Studies suggest that in many non-demented ALS patients there is impairment in attention, working memory, verbal fluency, and other frontal executive functions.

Researchers have questioned the relationship of speech dysfunction (i.e., severity of dysarthria) to the presence of cognitive impairment. Other studies have investigated whether cognitive dysfunction was associated with bulbar-onset illness. Strong and colleagues² found that patients with bulbar-onset disease showed greater impairment in working memory, problem-solving/cognitive flexibility, anomia, and recognition memory for words and faces, and their cognitive impairment was more progressive over time. However, they did not find a relationship with speech motor performance and intelligibility scores or respiration and cognition². Ringholz et al. found that patients with cognitive impairment (all degrees of impairment) were more likely to have motor programming difficulties, dysarthria, decreased phrase length and difficulty with word-finding¹. However, patients with bulbar-onset ALS did not differ from limb-onset patients in either level of impairment or pattern of cognitive performance¹.

SUBJECTS

- Recruited from The Baylor College of Medicine/The Methodist Hospital MDA/ALS Clinic
- All patients had a diagnosis of probable/definite ALS (El Escorial criteria)
- Males were significantly younger, more educated, and a higher VIQ, and were less dysarthric than females

Participant Demographics (N=175)

	Males Mean (SD)	Females Mean (SD)
Sample Size	112	63
Age	55.5 (13.4)	62.0 (15)*
Education	14.4 (3)	13.0 (3)**
VIQ-est.	103.12 (14.8)	94.42 (13.7)*
Dysarthria Sev-Appel (3,6,9,12)	5.25 (2.8)	6.6 (3.2)*
Dysarthria Sev-SpeechPath(1-7)	2.53 (1.5)	3.27 (1.8)

*p<.005, **p<.008

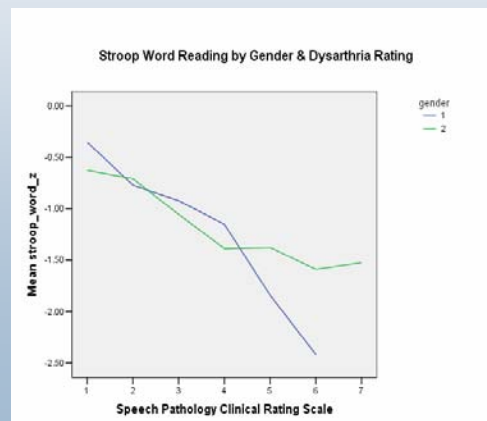
METHODS

ALS subjects completed an extensive battery of neuropsychological measures. Tests were chosen that assessed multiple areas of cognition, including:

- Modified (Satz-Mogel) version of the Wechsler Adult Intelligence Scale Revised (WAIS-R)
- American version of the National Adult Reading Test (AMNART)
- Stroop Color and Word Test
- Verbal Series Attention Test (VSAT)
- Verbal Fluency for letters (FAS) and Categories (animals)
- Logical Memory and Visual Reproduction subtests from the Wechsler Memory Scale-Revised (WMS-R)
- Symbol Digit Modalities Test (Oral and Written)
- Boston Naming Test
- Speech was assessed using the Appel ALS Rating Scale (ALS-Speech Score) and a speech pathologist's rating of dysarthria (Speech Pathology Clinical Dysarthria Scale)

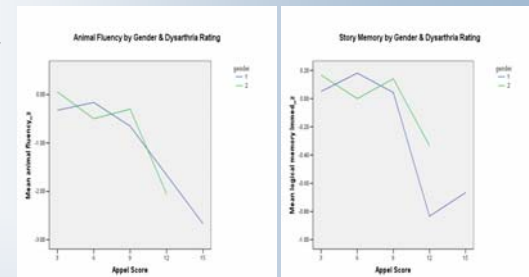
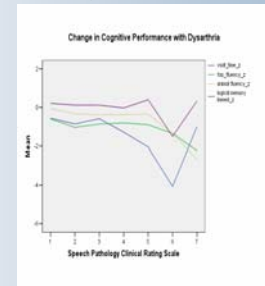
RESULTS

- There was a significant effect of Gender on cognitive performance across tests of intelligence, with males performing better than females, irrespective of dysarthria (including AMNART, est-VIQ, INFO, SIM, DSPAN, BD).
- There was a significant effect of Gender on Severity of Dysarthria, with women being more dysarthric than men (p= 0.003) (both rating scales). This probably relates to the higher rate of bulbar-onset in women.
- There was a significant overall effect of Dysarthria Severity on a few cognitive performances (see Figures):
STROOP-READING WORDS (p= 0.002, Appel; p= 0.001 Speech Path)
VSAT-TIME TO COMPLETION (p= 0.037, Appel; p= 0.017 Speech Path)
ANIMAL FLUENCY (p= 0.038, Appel; p= 0.076 Speech Path)
- There were no other significant effects of Dysarthria Severity on cognitive test performances, including tests of executive skills, attention, or learning and memory (See Figure).
- Post-hoc analyses indicated that the few significant differences were accounted for by the fact that older females tended to be more dysarthric.



RESULTS

- When the patients were divided into subgroups of those with normal to moderate dysarthria vs. those with moderately-severe to severe dysarthria, the patients in the severe dysarthria subgroup were found to perform significantly more poorly on:
ANIMAL FLUENCY (p= 0.003, Appel, n=3; p= 0.002 Sp.Path. N=4)
LEXICAL FLUENCY (p= 0.052, Appel, n=3; p= 0.029 Sp.Path. N=4)
STROOP-WORDS (p= 0.006, Appel, n=6; p= 0.002 Sp.Path. N=7)
VSAT-TIME (p= 0.068, Appel, n=5; p= 0.008 Sp.Path. N=6)
SYMBOL DIGIT-ORAL (p= 0.052, Appel, n=2; p= 0.000 Sp.Path. N=3)
STORY MEMORY-I (p= 0.044, Appel, n=7; p= 0.027 Sp.Path. N=8)
STORY MEMORY-II (p= 0.044 Sp.Path. n=8)
- The Speech Pathology Scale identified a broader range for dysarthria than the Appel Scale. Hence, it more sensitive to detecting effects on cognition.



CONCLUSIONS

There does not appear to be a significant linear effect of dysarthria severity on cognitive performance, but rather there appears to be a subgroup of patients with severe dysarthria who exhibit significantly poorer performance on some cognitive tests requiring verbal output, though not all tests assessing executive function. Thus, speech dysfunction manifesting as dysarthria is not solely responsible for the cognitive dysfunction seen in patients with ALS.

REFERENCES

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- Strong, M.J., Grace, G.M., Orange, J.B., Leeper, H.A., Menon, R.S., & Aere, C. (1999) A prospective study of cognitive impairment in ALS. *Neurology*, 53, 8, 1665-1670.