

Trajectory of spelling impairment in semantic variant of Primary Progressive Aphasia

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BACKGROUND

- Spelling difficulty is a known symptom of the semantic variant of Primary Progressive Aphasia (svPPA). Notably, svPPA is the only PPA variant with spelling impairment in the diagnostic framework (M.L. Gorno-Tempini et al., 2011).
- These impairments are typically described as surface dysgraphia, characterized by phonologically plausible spelling of irregular words (Josephy-Hernandez et al., 2023).
- While this pattern of surface dysgraphia is described in previous work (Shim et. al, 2012; Sepelyak et al., 2009), longitudinal characteristics of orthographic errors in svPPA have not been explored in depth.
- Here we aim to categorize the longitudinal pattern of spelling error types made by patients with svPPA using a novel orthographic error categorization system.
- Further, we explore relationships between the type of error patterns over time with overall language severity.
- We hypothesize that any preservation of irregularity in spelling patterns indicates a milder level of overall language impairment and slower progression due to maintenance of semantic memory.

METHODS

- We reviewed spelling errors that 20 participants with svPPA made on the Boston Diagnostic Aphasia Examination-3 (BDAE-3) (Goodglass et al., 1983) spelling task during language evaluations completed in our clinical research program.
- To categorize spelling patterns, we developed a novel error categorization system as follows:

Figure 1: Error Types

Error Type	Definition	Example
PARTIAL IRREGULARITY PRESERVATION (PI)	misspellings that retained some elements of the target word's atypical orthographic features	cough → caugh
REGULARIZATION (R)	phonologically plausible spellings reflecting classic surface dysgraphia	cough → coff
WHOLE WORD APPROXIMATIONS (WW)	real-word substitutions that were phonologically or orthographically similar to the target but were not plausible spellings of the target word	cough → coffee
OTHER (O)	responses with partial or minimal correspondence to the irregular target word, or no response	cough → couk
MIXED (M)	responses exhibiting characteristics of more than one error category	coffee+ yot
REGULAR WORD (RWE)	misspellings of words with regular, predictable spelling	apartment → apartment

- Spelling errors at three timepoints (baseline, midpoint, final) were categorized as above. Errors were independently coded by two speech-language pathologists; reliability analyses are ongoing.
- To explore potential relationships between spelling error trajectories and overall language severity, we analyzed baseline and annualized change in Progressive Aphasia Severity Scale Sum of Boxes (PASS-SB) scores, a rating of overall language impairment (Sapolsky et al., 2014). Annualized PASS-SB change was derived by averaging the rates of change across two consecutive visit intervals (T2-T1 and T3-T2), each calculated as score difference divided by inter-visit years.

METHODS (CONT.)

Table 1. Demographic and clinical characteristics of participants

Cohort	N = 20
Age (years)	64.5 ± 7.9
Sex (n %)	Male: 11 (55%) Female: 9 (45%)
Education (years)	16.9 ± 2.0
CDR Global (0-3)	0.475 ± 0.255
MoCA Score (0-30)	N = 10 19.7 ± 5.0
MMSE Score (0-30)	N = 9 23.4 ± 2.7

- Table 1. Values represent means and standard deviations
- CDR = Clinical Dementia Rating, MoCA = Montreal Cognitive Assessment, MMSE = Mini-Mental State Examination.
- Cognitive screening was assessed using either the MoCA or MMSE depending on clinical availability.

RESULTS

Figure 2: PASS-SB across error type trajectory groups by global aphasia severity



- Horizontal bars below each group indicate relative group size; numbers reflect participants per group.
- The predominance of regularization errors across the largest groups suggests that regularization-dominant spelling may be the most common error trajectory in svPPA.

Figure 4: PASS-SB across error type trajectory groups

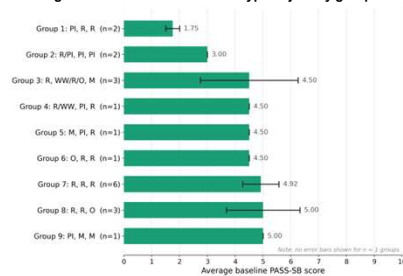
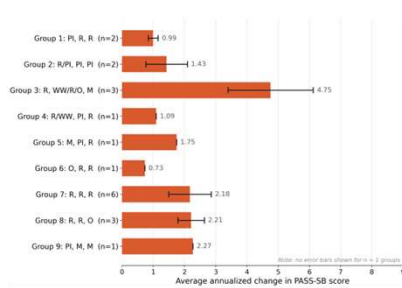


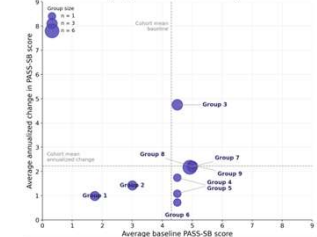
Figure 5: Annualized change in PASS-SB across error type trajectory groups



- Figures 4 and 5 show that a variance in baseline severity and rate of decline exist by error group type which suggests that spelling error pattern at initial presentation may serve as an early indicator of disease trajectory.

RESULTS (CONT.)

Figure 6: Clinical severity by baseline severity and annualized change



- Lower baseline PASS-SB scores and smaller annualized change in PASS-SB indicate milder impairment and slower progression.
- Group 3 has the highest baseline severity/fastest rate of decline and the most heterogeneity in error type and the absence of PI errors.
- Groups 1 and 2 cluster toward lower baseline severity/slower progression, have more homogeneity in error type, and include PI errors.

DISCUSSION

- Groups 7 and 8 (R,R,R and R,R,O) were the largest groups in our cohort, suggesting that regularization-dominant spelling may represent the most common error trajectory in svPPA. Compared to groups with PI errors at baseline, these participants showed higher levels of impairment as measured by PASS-SB scores and moderate rates of clinical progression, consistent with a steady decline in lexical orthographic access over time.
- In contrast, groups 1 (PI, R, R) and 2 (R/PI, PI, PI) have the two lowest baseline PASS-SB scores and two of the three lowest annualized change values (therefore the slowest progression). Both trajectory groups have partial irregularity (PI) errors at T1 and/or midpoint, either as the sole error type or co-occurring with regularization, potentially reflecting more preserved access to lexical orthographic knowledge early in their disease course. The presence of PI errors suggests at least some partial preservation of the semantic memory of how to spell these words rather than a complete loss of this knowledge.
- Group 3 (R, WW/R/O, M), with the more heterogeneous error trajectory and an absence of PI errors, is notable for having a higher baseline PASS-SB and by far the fastest progression based on annualized change. It may be that the increasing heterogeneity of error types (going from a single error type, to three co-occurring types, to a mixed-error pattern) is a signal of accelerating decline.

CONSIDERATIONS & FUTURE DIRECTIONS

- These are preliminary data for a descriptive study with a small sample size, including several single participant trajectory groups. The trends we describe here need to be replicated in larger samples.
- Characterizing longitudinal spelling error patterns in svPPA may ultimately help refine current models of orthographic impairment and guide the development of targeted assessment and monitoring approaches.
- It may be useful to generate a "reference guide" of errors made by participants with svPPA on a widely available spelling task, such as the one used here, for uniform classification of errors in the field. This will allow for a large data set of the classification of spelling errors in this patient population.

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