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Patterns of language decline in non-fluent primary progressive aphasia

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Abstract

Language samples collected yearly for up to 11 years post-onset of symptoms from four subjects presenting with non-fluent primary progressive aphasia (PPA) were analyzed and compared with samples collected from both non-brain-damaged subjects and those with agrammatic Broca's aphasia resulting from a single left-hemisphere stroke. Extensive analysis of lexical and morphosyntactic variables in these samples revealed two patterns of expressive language decline in the PPA subjects—one resembling that seen in our agrammatic aphasic subjects—i.e. impaired production of closed-class elements and loss of sentential structures governed by these elements—and the other characterized by advancing word-retrieval difficulties. These data are relevant for patient-management purposes and, in addition, they provide information relevant to language representation and organization.

Introduction

Primary progressive aphasia (PPA) is a clinical syndrome characterized by a gradual decline in aspects of language. The onset of the disorder is not marked by any acute neurological event; instead the disorder begins with subtle language difficulties (usually in the presenium) that often only the patient and his/her family members are able to detect. Originally described by Mesulam in 1982 and termed 'slowly progressive aphasia', the disorder is characterized by a gradual increase in both the number and severity of aphasic symptoms with a remarkable sparing of non-language cognitive abilities including memory, reasoning, insight, judgement, and comportment at least for a period of 2 years post-onset of symptoms (Weintraub *et al.* 1990, Weintraub and Mesulam 1992).

Although the language characteristics of individuals presenting with PPA have not been described in detail, deficits seen in these patients have been associated with those of aphasic individuals with acquired lesions. For example, fluent, non-fluent, and mixed fluent/non-fluent patterns have been reported. Both fluent and non-fluent cases of PPA often evince initial word-finding difficulties. However, as the disorder progresses in non-fluent cases, reduced phrase length and complexity, and telegraphic, agrammatic output (i.e. omission of obligatory morphology), are often noted, but auditory comprehension is relatively preserved (Duffy 1987, Duffy and Petersen 1992, Heath *et al.* 1983, Mesulam 1982, Weintraub *et al.* 1990). For

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example, Mesulam (1982) described five cases of PPA who initially presented with an anomic-like aphasia. These patients reported awareness of difficulty in word retrieval that worsened with time. Two of these subjects' aphasia (Mesulam's Subjects 1 and 3) became Broca-like as the disorder progressed, and the other three showed other patterns.

Although we estimate that at the present time approximately 100 patients with PPA have been described in the literature, none of these reports has provided more than general descriptions of the language decline seen in these patients. That is, detailed accounts of the decline of expressive language behaviours such as grammaticality of sentences, access to verbs and verb argument structure and grammatical morphology are not available. Such data are important both for patient-management reasons and for theoretical reasons. That is, this information is needed to aid in making accurate diagnoses, to make predictions and counsel patients about the course of language decline, and to plan intervention strategies. For example, it is likely that non-fluent PPA individuals demonstrate the same heterogeneity as seen in Broca's aphasic individuals with single-event vascular lesions and, therefore, different profiles of expressive language disruption and decline may be observed. Further, different treatments may be indicated at various stages of the disorder for patients with certain patterns of language decline. Indeed, treatment has recently been shown to affect the language ability of individuals with PPA (McNeil *et al.* 1995, Schneider *et al.* 1996); as we learn more about the ways in which language processes are affected during the course of decline we may be able to provide better strategies for intervention.

Detailed analysis of language decline is also of interest theoretically in that careful delineation of language structures and processes as they decline may provide insight into normal language representation and organization. For example, Basso *et al.* (1988) reported a category-specific semantic deficit in a patient with primary progressive aphasia that was very similar to that seen in some patients with focal lesions. That semantic categories fractionate as language declines in aphasia in a manner similar to that described in patients with focal lesions is informative in terms of organization of the lexicon. Such data are indeed provocative, and suggest that selective impairments of other language processes (e.g. access to verbs and verb argument structure or to classes of grammatical morphemes) also might be seen in PPA on close inspection of their language decline.

Thompson *et al.* (1994b) developed a detailed method for analysing language production that is motivated by linguistic theory (Chomsky 1986). The system details both lexical and morphosyntactic variables such as the proportions of certain sentence types and embeddings produced, verb type and verb argument structures produced, use of bound morphology, complexity and correctness of verb morphology, and ratios of nouns to verbs and open-class to closed-class words. Detailing these parameters of production has proven useful in studies of agrammatic aphasia. Research has shown, for example, that agrammatic aphasic subjects produce few grammatical sentences, more open-class as compared to closed-class words, and often more nouns than verbs (Goodglass 1976, Goodglass *et al.* 1993, Menn and Obler 1989, Saffran *et al.* 1989, Thompson *et al.* 1994b, 1995).

The purpose of the present study was to detail selected aspects of expressive language decline in four individuals with non-fluent PPA. Our PPA subjects were followed for up to 11 years post-onset of symptoms and, throughout its course, the aphasia and other behaviours were documented and analysed. In this study we undertook in-depth analysis of the expressive language samples collected. Profiles of our PPA patients were then compared to profiles of non-brain-damaged, non-language-impaired subjects and single left-hemisphere infarcted patients with moderately severe, agrammatic Broca's aphasia. This retrospective account represents an initial attempt to document the language decline of non-fluent PPA.

Method

Subjects

PPA subjects

Four subjects diagnosed with PPA, based on neurological and neuropsychological data, were included in the study. All subjects were right-handed and were native English speakers and minimally had 1 year of college education. They were between 40 and 61 years of age when aphasic symptoms were first clearly noted. Subjects 1 and 3 were females; Subjects 2 and 4 were males.

The Western Aphasia Battery (WAB, Kertesz 1982) or the Boston Diagnostic Aphasia Examination (BDAE, Goodglass and Kaplan 1983) and other language tests were administered and the Rating Scale Profile of Speech Characteristics from the BDAE was completed for all subjects (see Table 1). To establish reliability on the BDAE scale, two separate judges rated each subject on six of the seven parameters on the scale; point-to-point agreement between the two judges was calculated at 97%. On initial testing, deficits in melodic line and phrase length were not apparent for Subjects 1, 2, and 3; however, articulatory agility and grammatical form were considered to be mildly impaired. Subject 4 showed decreased melodic line, phrase length, articulatory agility and grammatical form even on initial testing. All subjects presented with mild deficits in naming, but spared auditory comprehension and repetition abilities.

In subsequent testing periods, productive language skills further declined in the face of sustained auditory comprehension ability. In the final test session Subjects 3 and 4 also showed declines in auditory comprehension—Subject 3 at 8 years post-onset of symptoms and Subject 4 at 6 years post-onset. All subjects received treatment for their aphasia at various times throughout the course of language decline; treatment was focused primarily on the use of compensatory strategies to augment communication.

Perceptual analysis of speech samples also was undertaken in order to evaluate the extent of dysarthria and motor speech programming impairments. Two judges rated 10 speech characteristics on a rating scale of 1–5 (1 = unimpaired; 5 = severely impaired). On initial testing, one of the subjects showed behaviours consistent with mild dysarthria (Subject 4); all subjects demonstrated mild articulatory deficits including infrequent phonemic repetition and transpositions. On subsequent samples collected midway through the data collection period, and during the final evaluation period, the subjects remained free of dysarthria (with the exception of Subject 4 whose dysarthria worsened over time). All subjects, however, showed declines in articulatory agility and speech rate (see Table 2).

Subjects also were administered a number of non-language neuropsychological tests including subtests of the Wechsler Adult Intelligence Scale-Revised (Wechsler 1981), the Wechsler Memory Scale (Wechsler and Stone 1945), the Hooper Visual

Table 1. Primary progressive aphasic (PPA) subjects' language test data (longitudinal)

	Subject 1 (A.H.)			Subject 2 (F.K.)					Subject 3 (M.K.)			Subject 4 (R.G.)				
Test period†	1	3	4	5	6	7	8	9	6	7	8	2	3	4	5	6
Boston Diagnostic Aphasia Examination (BDAE)																
Word Repetition (10)‡				8	9	8	5	5	10	9	2	9	9	8	7	7
Sentence Repetition (10)				10	9	9	4	5	12	6	0	7	10	4	6	1
Oral Word Reading (10)				10	9	10	9	8	10	10	0	9	9	8	7	3
Oral Sentence Reading (16)				8	9	7	5	6	7	3	0	6	7	4	3	1
Reading Comprehension (10)				10	9	9	9	9	10	10	8	8	10	8	8	7
Western Aphasia Battery (WAB)																
Fluency (10)	9	7	5													
Auditory Comprehension (10)	10	9	8·1													
Repetition (10)	7·8	7	4·2													
Naming (10)	10	8	7·6													
Reading (10)	10	10	10													
Aphasia Quotient (100)	93·6	81·5	65·8													
Cognitive Quotient (100)	95·6	89·2	82·3													
BDAE Rating Scale of Speech Characteristics																
Melodic Line	7	6	5	7		7		4	7	4	2	4		4		3
Phrase Length	7	6	4	7		7		4	7	4	1	4		4		2
Articulatory Agility	5·5	4·5	2	6		6		3	6	5	2	4		4		2
Grammatical Form	6	5	3	6·5		6·5		4	6	4	2	3		3		2
Paraphasia	7	7	7	7		7		7	7	7	—	7		7		—
Word Finding	4	5·5	6	4		4		5	4	6	7	5		5·5		7
Boston Naming Test (60)	56	48	38	48	49	43	32	32	57	48	5	49	49	43	37	24
Token Test																
Part V (23)	23	21	21	21	21	19	17	18	21	20	6	21	18	21	18	10

† Years post-onset of symptoms.
‡ Numbers in parentheses indicate maximum score.
Data for Subjects 2, 3 and 4 are from Weintraub *et al.* 1990.

Table 2. Perceptual analysis of dysarthria. Each speech dimension rated on a five-point scale (1 = normal; 5 = severe impairment)

Sample years post-symptoms	Subject 1 (A.H.)			Subject 2 (F.K.)			Subject 3 (M.K.)			Subject 4 (R.G.)		
	1	3	4	5	8	11	6	7	8	4	5	6
Consonant production	1	1	1	1	1	1	1	1	1	1	1	1
Articulatory agility	2	3	4	2	2	3	2	2	5	3	3	5
Resonance	1	1	1	1	1	1	1	1	1	1	1	1
Vocal Quality	1	1	1	1	1	1	1	1	1	1	1	1
Pitch	1	1	1	1	1	1	1	1	1	1	1	1
Loudness	1	1	1	1	1	1	1	1	1	1	1	1
Rate	1	2	3	1	1	2	1	2	3	2	3	4
Prolonged pauses	1	3	4	1	1	3	2	3	3	1	2	3
Intelligiblity	1	1	1	1	1	1	1	2	4	1	2	3

Organization Test (Hooper 1958), the Rey Auditory Verbal Learning Test (trial V) (Rey 1970), the Three Words–Three Shapes Test (Weintraub and Mesulam 1985), the Shipley–Hartford Institute of Living Scale (Shipley 1946) and other tests in the initial and in subsequent test sessions. Not all subjects were administered all tests on each test session, due to the severity of the aphasia and to time constraints; however, some of the tests were administered at each test interval. Results of this testing showed no decline in orientation, memory, visuospatial processing or reasoning over time. In addition, even on final testing, insight, judgement, and comportment were maintained. All subjects remained actively involved in activities in their communities and managed their own affairs throughout the period of language decline. See Weintraub *et al.* (1990) for detailed neuropsychological profiles of Subjects 2, 3, and 4 and Schneider *et al.* (1996) for descriptions of Subject 1.

On initial testing, elementary neurological examinations were largely unremarkable, with the exceptions that Subject 2 showed a mild right facial weakness which was not detected on subsequent examinations and Subject 3 showed a diminished swing of the right arm and bilateral posturing of her upper limbs on complex gait. Computerized tomography (CT) scans, magnetic resonance imaging scans (MRI), and electroencephalograms were unremarkable for Subjects 1, 2, and 4. Non-specific bifrontal atrophy, greater on the left side, was noted on both CT and MRI for Subject 3. Single-proton emission computerized tomography (SPECT), performed 2 years after initial examination for Subject 1, showed decreased perfusion in the left perisylvian region, and positron emission tomographic (PET) studies performed 2 years after initial examination for Subject 2 revealed reduced glucose metabolic activity in the left parietotemporal area. Elementary neurological examinations performed in final testing sessions were unchanged for Subjects 1, 2, and 3; Subject 4 showed bilateral dystonic posturing of the upper limbs on complex gait on final examination.

Comparison subject groups

Data from two comparison groups of subjects who participated in an earlier study (Thompson *et al.* 1994)—a group of five with agrammatic, Broca's aphasia resulting from a single, left-hemisphere stroke and a group of five non-brain-damaged subjects—were also used in the study. Subjects in the two comparison groups were matched for gender, age, and education. All subjects were monolingual, English-speaking, right-handed (with the exception of one aphasic subject who was left-handed), and passed a pure-tone audiometric screening at 40 dB HL, ANSI:1969 in at least one ear.

The aphasic subjects were at least 2 years post-stroke; WAB Aphasia Quotients ranged from 62·4 to 77·6. Analysis of the production patterns of these subjects showed behaviours consistent with a diagnosis of agrammatism (see agrammatic patient profiles presented by Saffran *et al.* 1989 and Menn and Obler 1989). Utterance length was reduced, and less than 50% of sentences were grammatical; grammatical morphemes were frequently missing, as were obligatory verb argument structures. They produced a greater proportion of open as compared to closed class words (*M* open:closed class ratio = 1·56) and within the category of open-class words, a greater proportion of nouns as compared to verbs was produced (*M* = 1·48). Auditory comprehension was relatively intact for all subjects (WAB scores ranged from 7·9 to 9·0 based on a total possible score of 10). Additional testing of auditory comprehension indicated good lexical (single-word) and active sentence comprehension. Grammaticality judgement was also good, but comprehension of non-canonical sentences was impaired.

Procedure

Discourse samples—either narrative or conversational—were collected for the subjects with PPA at 1-year intervals over a 3–7-year period and analysed retrospectively. All samples were collected by one of the authors or by an ASHA-certified speech–language pathologist. Samples for Subject 1—collected in Thompson's research laboratory—were derived by asking her to tell the story of Cinderella using methods described by Saffran *et al.* (1989). Discourse samples for the other PPA subjects (Subjects 2, 3, and 4)—collected in Weintraub and Mesulam's laboratory—were derived from patient/clinician conversational samples concerned with history of illness and related problems, and description of the cookie theft picture from the BDAE (Goodglass and Kaplan 1983). A total of four samples was collected for Subject 1, seven samples were collected for Subject 2, three for Subject 3, and three for Subject 4 ($n = 17$). Conversational discourse samples were also collected from the subjects in the two comparison groups; these subjects were asked to watch a 5-minute TV news segment and then to discuss it (or talk about anything else that they wished) for approximately 10 minutes. It is important to point out here that research has indicated that the linguistic patterns seen in both agrammatic aphasic subjects and in non-brain-damaged subjects is stable across discourse conditions (Thompson *et al.* 1994b, 1995, Rochon *et al.* 1994). Thompson *et al.* (1995) found no statistically significant differences between narrative (i.e. Cinderella stories) and conversational discourse tasks (i.e. discussion of TV news segments) on the proportion of complex sentences produced, noun:verb ratio, open-class:closed-class ratio and other variables for either population. For example, the mean proportion of complex sentences produced by the aphasic subjects was 0·20 (SD = 0·18) and 0·27 (SD = 0·20) for the narrative and conversational discourse conditions, respectively. For the normal subjects the two means were 0·57 (SD = 0·17) and 0·58 (SD = 0·09) (Thompson *et al.* 1995, p. 126). Thompson *et al.* (1996) have also found stable grammatical behaviours across testing periods (with up to 2-week intervals between samples) on both narrative and conversational conditions in agrammatic and non-brain-damaged subjects.

All samples were audiotaped, transcribed and entered into a computer for analysis. Samples were segmented into utterances based on syntactic, prosodic, and semantic criteria. A minimum of 42 propositional utterances was included in each sample. Each utterance was coded linguistically using the method developed by Thompson *et al.* (1994b). This involved coding sentences for grammaticality and complexity, lexical items by category, and verbs by type and by argument structure characteristics. The correctness and complexity of verb morphology was also coded, and a verb morphology index (VMI) was assigned to each verb phrase. This index was calculated by giving a point each for the verb stem, affixes, modals, auxiliaries, realization of tense and agreement on auxiliaries, and infinitival 'to'. Clitics (contracted auxiliaries) and bound morphemes occurring on nouns were also coded for accuracy.

Coded samples were then analysed using the Systematic Analysis of Language Transcripts software program (MacSALT: Miller and Chapman 1992). This program automatically calculates the mean length of utterance and provides frequency counts of each code. The proportion of grammatical sentences produced, the proportion of simple and complex sentences, mean embeddings, the proportion of verbs produced with correct morphology, and the mean complexity of verb morphology (mean VMI) were computed for each sample. The proportion of verbs produced by type and the proportion produced with correct argument structure was also computed. We also computed the proportion of specific verb arguments such as Agent, Theme, and Goal that were produced in the utterances of our subjects. Finally, ratios of open-class to closed-class words and nouns to verbs were computed.

Reliability

All language samples were transcribed by two of the authors in order to determine reliability of data entry and utterance segmentation. Overall point-to-point agreement on data entry was 98% and agreement on utterance segmentation was 96%. Disagreements were resolved by discussion between the transcriptionists and (when necessary) by re-listening to the samples in question.

Two of the authors also independently coded all language samples, and inter-coder point-to-point reliability was calculated for each variable (i.e. sentence grammaticality codes, verb codes, etc.). Overall agreement ranged from 87% to 90% with an overall mean of 88·5%.

Results

Data derived from the discourse samples of each of our PPA subjects are shown in Tables 3–6. These data show progressive decline in all language variables examined across all subjects.

One of the primary purposes of this study was to compare the language patterns noted throughout the progression of PPA to language patterns seen in our focal-lesion agrammatic aphasic subjects and in our non-brain-damaged subjects. In the following sections we present these comparisons for the following selected

Table 3. **Lexical and morphosyntactic variables in discourse samples collected for Subject 1 (A.H.)**

Years post-onset of symptoms	Sample 1 Year 1	Sample 2 Year 2	Sample 3 Year 3	Sample 4 Year 4
Sentence variables				
MLU	6·50	5·76	7·20	4·72
Total number of utterances	30	55	44	46
Grammatical sentences (proportion)	0·32	0·23	0·06	0·07
Simple sentences	0·56	0·89	0·81	0·93
Complex sentences	0·44	0·11	0·19	0·07
Mean embeddings	0·48	0·11	0·22	0·11
Mean verb morphology index	2·30	2·14	2·00	1·90
Correct verb morphology	0·80	0·73	0·50	0·43
Verb types				
Total verbs (proportion produced (proportion correct))	40	58	60	30
Obligatory one-place	0·00	0·00	0·00	0·00
Obligatory two-place	0·08 (0·33)	0·05 (0·33)	0·02 (0·00)	0·03 (1·00)
Obligatory three-place	0·00	0·05 (0·33)	0·00	0·00
Optional two-place	0·35 (0·79)	0·50 (0·52)	0·41 (0·45)	0·60 (0·33)
Optional three-place	0·13 (0·40)	0·10 (0·33)	0·15 (0·22)	0·20 (0·50)
Copula	0·10 (0·57)	0·03 (0·00)	0·02 (0·00)	0·00
Complement	0·34 (0·00)	0·27	0·40 (0·25)	0·17 (0·00)
Verbs with simple argument structure	0·50 (0·80)	0·75 (0·55)	0·90 (0·73)	0·80 (0·50)
Verbs with complex argument structure	0·50 (0·80)	0·25 (0·25)	0·10 (0·00)	0·20 (0·00)
Total verbs produced with correct argument structure	0·65	0·44	0·42	0·4
Argument structure data (proportion correct)				
Agents	0·76 ($n = 34$)	0·57 ($n = 58$)	0·57 ($n = 60$)	0·52 ($n = 33$)
Themes	0·83 ($n = 30$)	0·79 ($n = 43$)	0·69 ($n = 36$)	0·72 ($n = 25$)
Goals	0·67 ($n = 3$)	0·17 ($n = 6$)	0·29 ($n = 7$)	0·50 ($n = 2$)
Sentential complements	0·63 ($n = 8$)	0·25 ($n = 4$)	0·60 ($n = 5$)	0·50 ($n = 2$)
Predication phrases	0·75 ($n = 8$)	0·67 ($n = 3$)	0·00 ($n = 1$)	0·33 ($n = 3$)
Phrasal adjuncts	0·25 ($n = 4$)	0·77 ($n = 13$)	0·27 ($n = 12$)	0·45 ($n = 7$)

Lexical variables				
Total words	193	306	317	206
Nouns (proportion)	0·22	0·29	0·32	0·45
Verbs (proportion)	0·21	0·19	0·19	0·15
Adjectives (proportion)	0·08	0·08	0·06	0·06
Adverbs (proportion)	0·03	0·02	0·02	0·02
Noun/verb ratio	1·08	1·55	1·70	3·06
Open-class words	0·53	0·58	0·60	0·68
Closed-class words	0·47	0·43	0·40	0·32
Open-class/closed-class ratio	1·14	1·35	1·48	2·12
Bound morpheme data				
Clitics (contracted auxiliaries)				
Number produced (number omitted)	3 (0)	1 (0)	0 (0)	1 (4)
Verb morphology				
Number produced (number omitted)	7 (0)	8 (4)	9 (8)	12 (1)
Noun morphology				
Number produced (number omitted)	6 (0)	7 (0)	20 (1)	4 (3)

Table 4. Lexical and morphosyntactic variables in discourse samples collected for Subject 2 (F.K.)

ırs post-onset of symptoms	Sample 1 Year 5	Sample 2 Year 6	Sample 3 Year 7	Sample 4 Year 8	Sample 5 Year 9	Sample 6 Year 10	Sample 7 Year 11
ıtence variables							
.U	5·90	6·86	5·54	4·81	3·69	3·24	2·65
:al utterances	82	123	163	113	101	123	182
ımmatical sentences (proportion)	0·61	0·68	0·62	0·64	0·43	0·48	0·41
›imple sentences	0·49	0·68	0·68	0·66	0·68	0·70	0·90
ᑐomplex sentences	0·51	0·32	0·32	0·34	0·32	0·30	0·10
an embeddings	0·77	0·42	0·35	0·55	0·34	0·25	0·08
an verb morphology dex	2·24	2·40	2·42	2·45	2·33	2·23	1·98
rrect verb .orphology	0·99	0·98	0·99	0·94	0·87	0·86	0·87
rb types							
tal verbs	91	147	151	91	61	57	54
(proportion produced (proportion correct))							
Obligatory one-place	0·08 (1·00)	0·03 (0·75)	0·05 (1·00)	0·03 (1·00)	0·10 (0·50)	0·02 (1·00)	0·04 (1·00)
Obligatory two-place	0·02 (1·00)	0·10 (0·93)	0·12 (0·83)	0·15 (0·93)	0·03 (0·50)	0·07 (0·75)	0·06 (0·67)
Obligatory three-place	0·00	0·03 (0·25)	0·03 (0·50)	0·01 (0·00)	0·03 (0·00)	0·04 (0·50)	0·00
Optional two-place	0·14 (0·92)	0·20 (0·90)	0·19 (0·86)	0·14 (0·85)	0·23 (0·71)	0·26 (0·60)	0·31 (0·59)
Optional three-place	0·01 (1·00)	0·03 (0·75)	0·03 (0·80)	0·03 (1·00)	0·03 (1·00)	0·02 (1·00)	0·02 (0·00)
Copula	0·24 (0·80)	0·20 (0·97)	0·15 (0·86)	0·26 (0·79)	0·18 (0·64)	0·14 (0·88)	0·09 (0·20)
Complement	0·51 (0·48)	0·41 (0·30)	0·40 (0·77)	0·38 (0·45)	0·40 (0·65)	0·45 (0·62)	0·48 (0·88)
rbs with simple ɾgument structure	0·59 (0·86)	0·70 (0·91)	0·77 (0·70)	0·61 (1·00)	0·78 (0·73)	0·65 (0·81)	0·88 (0·57)
:rbs with complex rgument structure	0·41 (0·74)	0·30 (0·81)	0·23 (0·92)	0·39 (0·54)	0·22 (0·60)	0·35 (0·78)	0·12 (0·00)
›tal verbs produced vith correct argument tructure	0·87	0·89	0·81	0·82	0·67	0·75	0·52

Argument structure data (proportion correct)							
Agents	0·99 (*n* = 67)	0·95 (*n* = 112)	0·95 (*n* = 128)	0·97 (*n* = 65)	0·82 (*n* = 51)	0·88 (*n* = 51)	0·88 (*n* = 48)
Themes	0·91 (*n* = 64)	0·97 (*n* = 103)	0·82 (*n* = 113)	0·91 (*n* = 68)	0·86 (*n* = 42)	0·89 (*n* = 38)	0·54 (*n* = 37)
Goals	1·00 (*n* = 4)	0·70 (*n* = 10)	0·78 (*n* = 9)	0·50 (*n* = 2)	0·83 (*n* = 6)	0·67 (*n* = 3)	1·00 (*n* = 1)
Sentential complement	0·86 (*n* = 22)	0·85 (*n* = 20)	0·93 (*n* = 14)	0·71 (*n* = 17)	0·57 (*n* = 7)	0·78 (*n* = 9)	0·00 (*n* = 3)
Predication phrases	0·82 (*n* = 22)	0·97 (*n* = 36)	0·88 (*n* = 26)	0·87 (*n* = 23)	1·00 (*n* = 7)	0·86 (*n* = 7)	0·50 (*n* = 8)
Phrasal adjuncts	0·85 (*n* = 20)	0·64 (*n* = 28)	0·80 (*n* = 25)	0·74 (*n* = 19)	0·67 (*n* = 9)	1·00 (*n* = 1)	0·38 (*n* = 8)
Lexical variables							
Total words	431	726	766	439	274	229	257
Nouns (proportion)	0·16	0·14	0·16	0·15	0·18	0·14	0·16
Verbs (proportion)	0·21	0·20	0·20	0·21	0·22	0·25	0·21
Adjectives (proportion)	0·08	0·08	0·06	0·05	0·04	0·04	0·04
Adverbs (proportion)	0·06	0·06	0·06	0·08	0·09	0·07	0·07
Noun/verb ratio	0·75	0·71	0·81	0·74	0·81	0·58	0·76
Open class	0·51	0·48	0·47	0·48	0·53	0·50	0·48
Open-class words	0·51	0·48	0·47	0·48	0·53	0·50	0·48
Closed-class words ratio	0·49	0·52	0·53	0·52	0·47	0·50	0·52
Bound morpheme data							
Clitics (contracted auxiliaries)							
Number produced (number omitted)	14 (0)	18 (0)	22 (0)	·24 (0)	6 (1)	6 (0)	6 (1·00)
Verb morphology							
Number produced (number omitted)	23 (2)	32 (0)	38 (0)	21 (0)	13 (1)	6 (0)	4 (0)
Noun morphology							
Number produced (number omitted)	11 (0)	11 (0)	16 (0)	18 (0)	7 (0)	2 (0)	4 (0)

Table 5. Lexical and morphosyntactic variables in discourse samples collected for Subject 3 (M.K.)

ırs post-onset of symptoms	Sample 1 Year 6	Sample 2 Year 7	Sample 3 Year 8
ıtence variables			
ᴶU	5·59	4·94	3·25
tal utterances	89	42	50
ammatical sentences (proportion)	0·31	0·12	0·00
Simple sentences	0·91	0·82	0·80
Complex sentences	0·09	0·18	0·20
:an embeddings	0·09	0·18	0·20
:an verb morphology index	2·26	267	1·80
:rect verb morphology	0·72	0·44	0·40
rb types	37	17	5
ıtal verbs (proportion produced (proportion correct))			
Obligatory one-place	0·03 (1·00)	0·06 (0·00)	0·00
Obligatory two-place	0·08 (0·67)	0·24 (0·75)	0·20 (1·00)
Optional two-place	0·05 (0·50)	0·18 (0·67)	0·20 (0·06)
Optional three-place	0·05 (0·50)	0·12 (0·00)	0·00
Copula	0·30 (0·36)	0·12 (0·00)	0·00
Complement	0·52 (0·41)	0·49 (0·25)	0·60 (0·00)
ɔtal verbs produced with correct argument structure	0·43	0·35	0·20
rgument structure data (proportion correct)			
Agents	0·50 ($n = 26$)	0·63 ($n = 16$)	0·17 ($n = 6$)
Themes	0·83 ($n = 42$)	0·75 ($n = 20$)	0·50 ($n = 4$)
Goals	1·00 ($n = 3$)	0·00 ($n = 2$)	($n = 0$)
Sentential complements	0·00 ($n = 1$)	0·50 ($n = 2$)	($n = 0$)
Predication phrases	0·13 ($n = 8$)	0·33 ($n = 3$)	($n = 0$)
Phrasal adjuncts	0·71 ($n = 24$)	0·57 ($n = 7$)	0·00 ($n = 1$)

Lexical variables			
Total words	387	196	165
Nouns (proportion)	0·36	0·35	0·42
Verbs (proportion)	0·10	0·09	0·03
Adjectives (proportion)	0·14	0·13	0·22
Adverbs (proportion)	0·01	0·01	0·00
Noun/verb ratio	3·73	4·00	13·80
Open-class words	0·60	0·57	0·67
Closed-class words	0·40	0·43	0·33
Open/closed class ratio	1·51	1·33	2·00
Bound morpheme data			
Clitics (contracted auxiliaries)			
Number produced (number omitted)	4 (0)	1 (3)	0 (1)
Verb morphology			
Number produced (number omitted)	10 (6)	9 (3)	7 (0)
Noun morphology			
Number produced (number omitted)	22 (3)	13 (4)	11 (0)

Table 6. Lexical and morphosyntactic variables in discourse samples collected for Subject 4 (R.G.)

Years post-diagnosis	Sample 1 Year 4	Sample 2 Year 5	Sample 3 Year 6
Sentence variables			
MLU	3·76	3·75	1·87
Total utterances	42	44	62
Grammatical sentences (proportion)	0·00	0·07	0·00
Simple sentences	1·00	1·00	1·00
Complex sentences	0·00	0·00	0·00
Mean embeddings	0·00	0·00	0·00
Mean verb morphology index	2·10	1·94	1·43
Correct verb morphology index	0·50	0·19	0·14
Verb types			
Total verbs (proportion produced (proportion correct))	10	16	7
Obligatory one-place	0·10 (1·00)	0·13 (0·00)	0·00
Optional two-place	0·40 (0·25)	0·50 (0·00)	0·29 (0·50)
Optional three-place	0·30 (0·00)	0·13 (0·00)	0·57 (0·25)
Copula	0·10 (0·00)	0·25 (0·00)	0·14 (0·00)
Complement	0·10 (0·00)	0·00	0·00
Total verbs produced with correct argument structure	0·20	0·00	0·29
Argument structure data (proportion correct)			
Agents	0·30 ($n = 10$)	0·00 ($n = 12$)	0·33 ($n = 6$)
Themes	0·78 ($n = 9$)	0·36 ($n = 11$)	0·00 ($n = 1$)
Goals	($n = 0$)	($n = 0$)	($n = 0$)
Sentential	($n = 0$)	($n = 0$)	($n = 0$)
Predication phrases	1·00 ($n = 1$)	0·50 ($n = 4$)	0·00 ($n = 1$)
Phrasal adjuncts	0·33 ($n = 3$)	0·00 ($n = 6$)	0·00 ($n = 1$)

Lexical variables			
Total words	90	75	37
Nouns (proportion)	0·49	0·40	0·38
Verbs (proportion)	0·11	0·21	0·19
Adjectives (proportion)	0·10	0·19	0·24
Adverbs (proportion)	0·01	0·01	0·00
Noun/verb ratio	4·40	1·88	2·00
Open-class words	0·71	0·81	0·84
Closed-class words	0·29	0·19	0·16
Open class/closed class ratio	2·46	4·36	5·17
Bound morpheme data			
Clitics (contracted auxiliaries)			
Number produced (number omitted)	0 (1)	0 (2)	0 (0)
Verb morphology			
Number produced (number omitted)	3 (3)	5 (1)	2 (1)
Noun morphology			
Number produced (number omitted)	3 (0)	3 (0)	3 (0)

variables: mean length of utterance, the proportion of grammatical sentences, the proportion of verbs produced with correct morphology, the complexity of verb morphology (VMI), the proportion of verbs produced with correct argument structure, open-class to closed-class word ratio, and noun to verb ratio. For each of these variables we compare the PPA subjects' performance to the mean performance (±1 standard deviation) for our agrammatic Broca's aphasic and non-brain-damaged subjects.[1]

Mean length of utterance (MLU)

Similar patterns of decline in MLU were noted across subjects, although Subject 4's MLU declined before that of the other subjects. As shown in Figure 1, all of the PPA subjects presented with MLU below the range of our non-brain-damaged subjects even on initial testing, and all showed performance within the range of our agrammatic aphasic subjects at some point in the course of disease progression. MLU for Subject 1 was 6·50 at 2 years post-diagnosis; by year 4 it had declined to 4·72. At 5 years post-diagnosis, Subject 2 evinced an MLU of 5·90; by year 9 it was approximately at the mean of our agrammatic aphasic subjects, and by year 11 it had declined to below the agrammatic mean to 2·65. Very similar patterns were noted for Subjects 3 and 4, although Subjects 4's MLU was poorer on initial testing and it remained within the range of our agrammatic subjects on all testings.

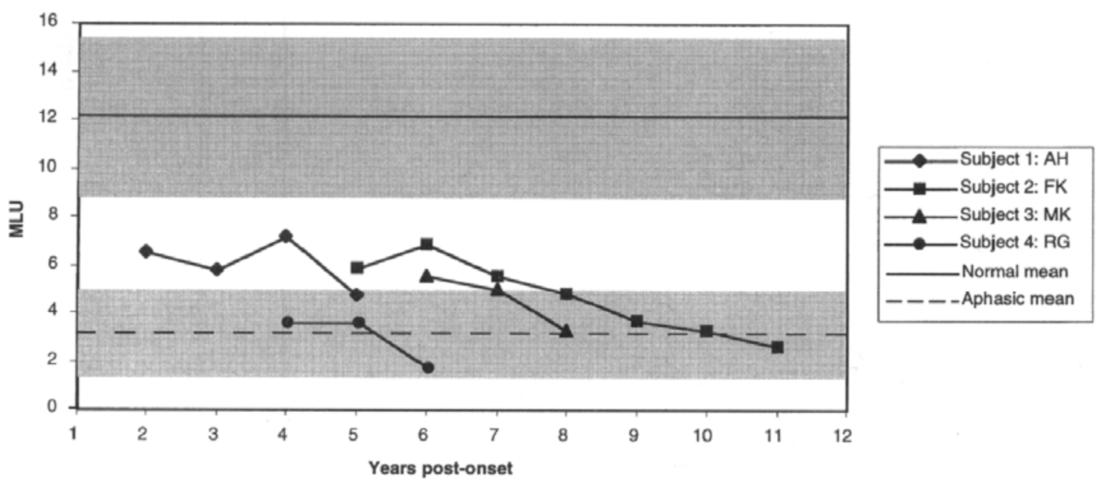

Figure 1. Decline of mean length of utterance (MLU) seen in primary progressive aphasic (PPA) subjects over time—years post-onset of diagnosis. The mean values (±1 standard deviation) for non-brain-damaged and agrammatic aphasic comparison groups are provided ($M = 12{\cdot}18$; $SD = 3{\cdot}24$ and $M = 3{\cdot}19$; $SD = 1{\cdot}72$, respectively).

Proportion of grammatical sentences

Figure 2 depicts the proportion of grammatical sentences produced by both of our comparison groups and by our PPA subjects. As can be seen, a low proportion of grammatical sentences was noted in the initial samples derived from three of the subjects with PPA, Subjects 1, 3, and 4. Subject 1 showed performance within the

[1] We note that the language patterns of our agrammatic Broca's aphasic subjects are quite similar to those derived from other in-depth studies of agrammatic aphasia. See for example studies by Saffran *et al.* (1989) and by Menn and Obler (1989). Similarly, the data from our non-brain-damaged subjects matches those reported in Saffran *et al.* study, and by others.

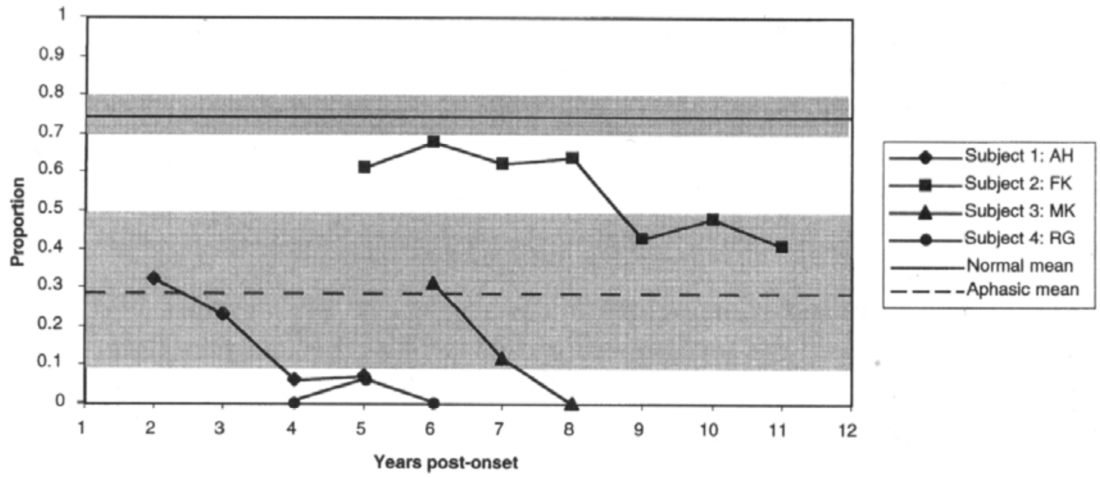

Figure 2. Proportion of grammatical sentences produced over time (by years post-diagnosis) by subjects with primary progressive aphasia (PPA). The mean values for non-brain-damaged and agrammatic aphasic comparison groups are provided ($M = 9{\cdot}75$; SD = 0·05 and $M = 0{\cdot}28$; SD = 0·20, respectively).

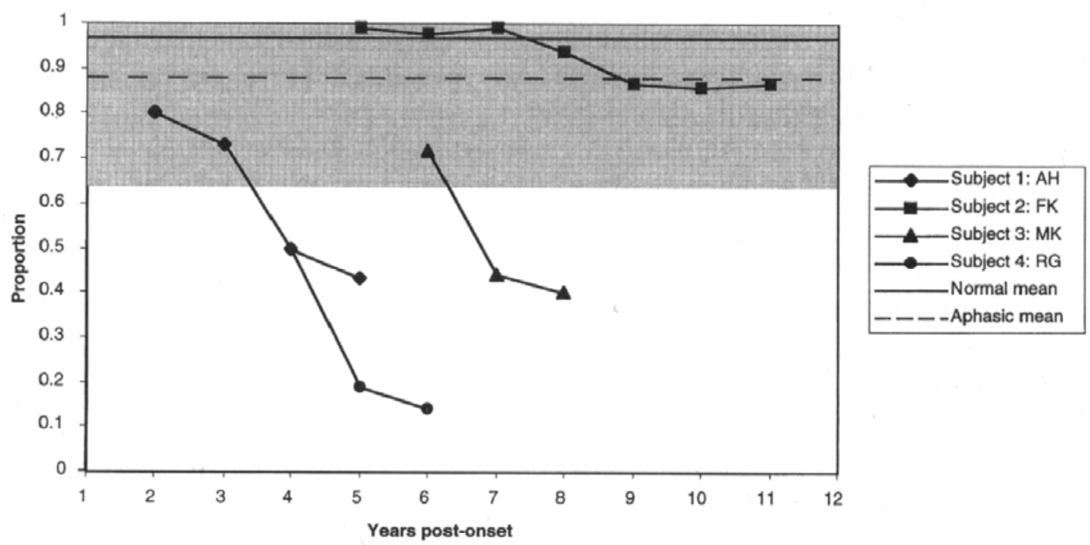

Figure 3. Proportion of verbs produced with correct morphology over time (by years post-diagnosis) for the subjects with primary progressive aphasia (PPA). The mean values for non-brain-damaged and agrammatic aphasic subjects are provided for comparison ($M = 0{\cdot}97$; SD = 0·0 and $M = 0{\cdot}88$; SD = 0·25, respectively).

range of our agrammatic comparison group (0·32) at 2 years post-diagnosis; however, by year 4 she produced few grammatical sentences and her performance was below the range of our focal-lesioned subjects. Similarly, by year 4 post-onset of symptoms, Subject 4 produced few grammatical sentences—a pattern which persisted throughout the data-collection period for this subject. The decline of grammatical sentences was somewhat slower for Subject 3; her performance fell at the mean of our agrammatic aphasic subjects at 6 years post-onset of symptoms, and did not decline to levels below that seen in the focal-lesioned agrammatic subjects until 8 years post-onset. For all three of these subjects the proportion of grammatical sentences declined to values at or near zero (range of 0·00–0·07).

Subject 2, however, showed a somewhat different course. He produced a higher proportion of grammatical sentences in the first sample (5 years post-diagnosis) than did the other PPA subjects, and up to year 8 post-onset of symptoms his performance was only slightly below the range of our non-brain-damaged subjects.

Further, he did not show the marked decline that was noted in the other PPA subjects; even at year 11 post-symptoms, sentence grammaticality was approximately one standard deviation above the mean of our agrammatic aphasic subjects.

Verb morphology

Accuracy

Bound and free-standing morphemes occurring in the verb group were analysed both for complexity and for accuracy. Figure 3 depicts the accuracy data. As can be seen, the PPA subjects once again showed two patterns of performance. Subject 1 showed performance similar to that of our agrammatic subjects at 2 and 3 years post-onset of symptoms; however, she declined to even lower levels by year 4. Subject 4's performance at year 4 was consistent with that of Subject 1's at year 4, and it continued to decline in subsequent years. Subject 3 showed a similar pattern of decline, although once again her decline occurred much later post-onset of symptoms.

Subject 2 showed a different pattern. The proportion of verbs produced with correct morphology remained within the range of our non-brain-damaged subjects (0·99–0·87) until 9 years post-symptoms; even at 11 years post-onset his performance remained less impaired than the other PPA subjects' performances.

Complexity (VMI)

Verb morphology indices are shown in Figure 4. All of our PPA subjects had reduced complexity of verbal morphology and demonstrated performance levels similar to our agrammatic aphasic subjects. Gradual declines were apparent for Subjects 1 and 4, and Subject 3's performance declined similarly, but at a later time. Subject 2 demonstrated higher VMI levels than the other subjects with performance above the range of our agrammatic subjects even at 8 years post-onset. Complexity of verb morphology declined, however, to levels similar to the other PPA subjects on later samples.

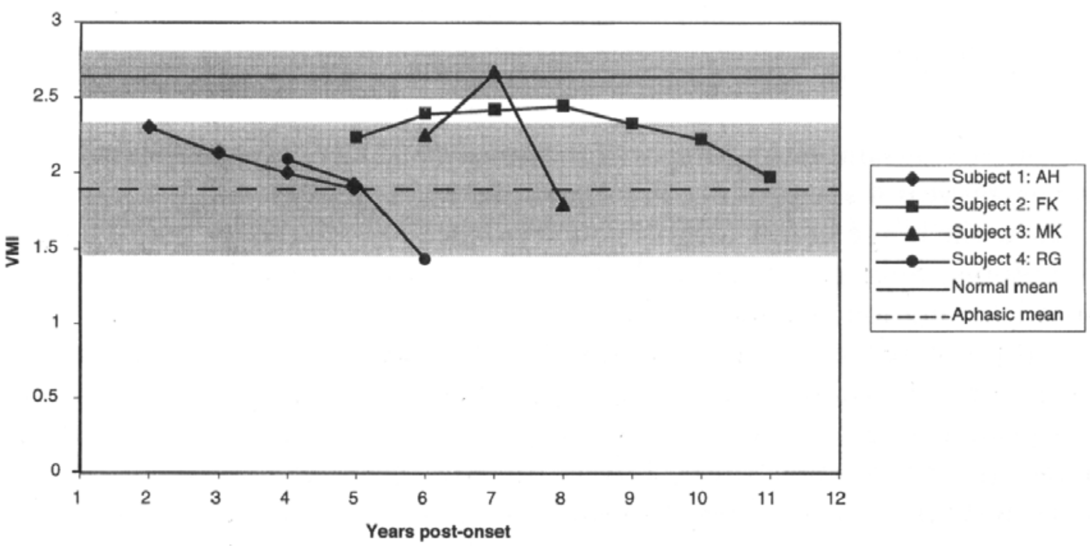

Figure 4. Complexity of verb morphology produced over time (by years post-diagnosis) for the subjects with primary progressive aphasia (PPA). Mean values for non-brain-damaged and agrammatic aphasic subjects are provided for comparison ($M = 2{\cdot}65$; SD $= 0{\cdot}15$ and $M = 1{\cdot}90$; SD $= 0{\cdot}42$, respectively).

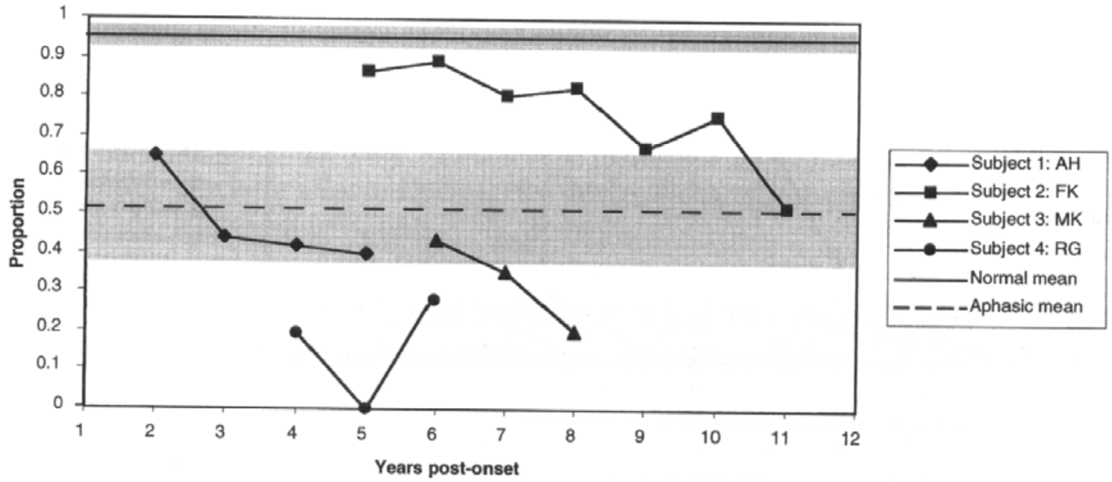

Figure 5. Proportion of verbs produced with correct argument structure over time (by years post-diagnosis) by the subjects with primary progressive aphasia (PPA). Mean values for non-brian-damaged and agrammatic aphasic comparison groups are provided ($M = 0{\cdot}95$; SD $= 0{\cdot}03$ and $M = 0{\cdot}51$; SD $= 0{\cdot}14$, respectively).

Proportion of verbs produced with correct argument structure

Figure 5 depicts the proportion of verbs produced with correct argument structure. PPA Subjects 1 and 3 performed similarly to the agrammatic subjects, although Subject 3's performance declined to lower levels at the final evaluation. Subject 4 showed very poor performance, producing verb argument structures more poorly than the other subjects at an earlier point post-onset of symptoms. Interestingly, as we discuss later, this subject also produced fewer verbs than the other subjects—a factor which also influenced the grammaticality of his sentences.

In contrast, Subject 2 once again showed performance far superior to our other PPA subjects. Although his performance was poorer than that of the non-brain-damaged subjects, he produced a greater proportion of verb argument structures correctly than did our agrammatic comparison subjects. Until his final evaluation, at year 11, he produced between 70% and 90% of verbs with their correct arguments.

Open- to closed-class word ratio

Data depicting open : closed-class ratios for our subjects are shown in Figure 6. The non-brain-damaged subjects produced approximately equal numbers of open- and closed-class words with mean ratio of 0·93 (SD = 0·11). The agrammatic aphasic comparison subjects produced an average open : closed class ratio of 1·56 (SD = 0·39), indicating a typical pattern seen in agrammatic aphasia. That is, they produced more open-class than closed-class words.

Like the agrammatic subjects, Subjects 1, 3 and 4 showed a greater number of open-class as compared to closed-class words, and the proportion of closed-class words declined markedly with the progression of the disorder. Ratios increased over time from 1·14 to 2·12 for Subject 1 and from 1·51 to 2·00 for Subject 3. Once again, however, Subject 3 showed decline later in the course of disease progression. Subject 4 showed severe impairment by 4 years post-symptoms with ratios further declining from 2·46 to 5·17 by year 6. In contrast, Subject 2 demonstrated open : closed-class ratios similar to that for the normal subjects and no change was observed over time (1·03 to 0·92).

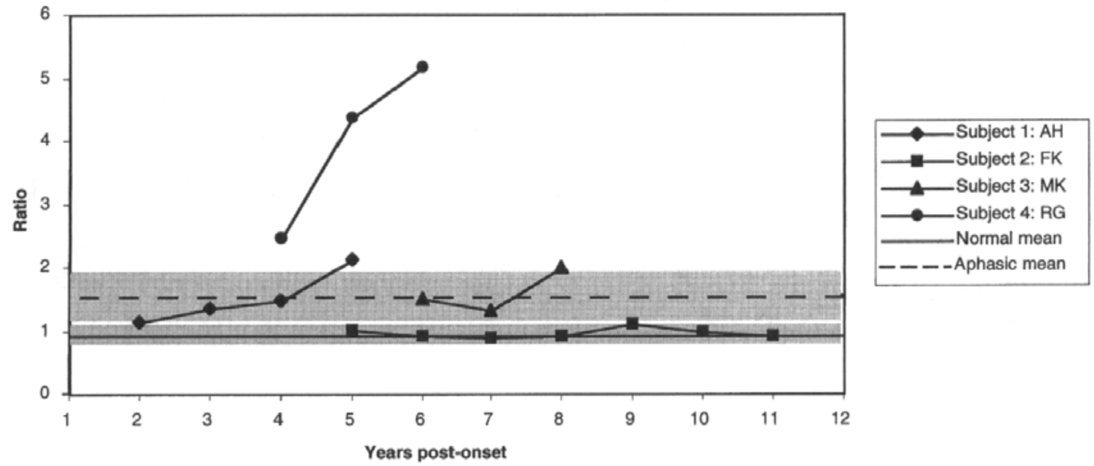

Figure 6. The ratio of open-class to closed-class words produced over time (by years post-diagnosis) by the primary progressive aphasic (PPA) subjects. Mean values for non-brain-damaged and agrammatic, Broca's aphasic subjects are provided for comparison ($0{\cdot}93 \pm 0{\cdot}11$ and $1{\cdot}56 \pm 0{\cdot}39$, respectively).

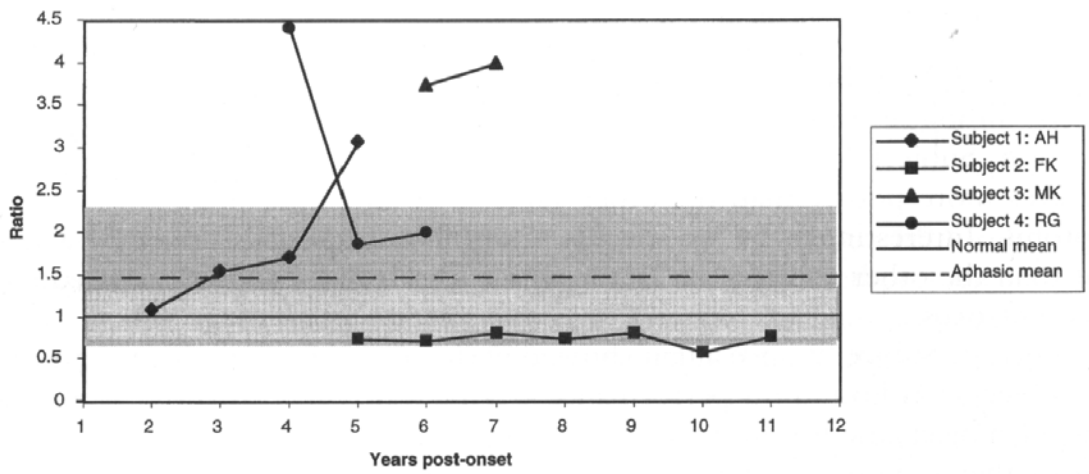

Figure 7. The ratio of nouns to verbs produced over time (by years post-onset of symptoms) by PPA subjects. The mean values for non-brain-damaged and agrammatic aphasic subjects are provided for comparison ($M = 1{\cdot}01$; SD $= 0{\cdot}3$ and $M = 1{\cdot}48$; SD $= 0{\cdot}80$, respectively). The third data point for Subject 3 (ratio $= 13{\cdot}80$) is omitted from the chart.

Noun to verb ratio

Noun : verb ratios derived for our subjects are shown in Figure 7. The non-brain-damaged subjects produced about equal numbers of nouns and verbs, with the mean ratio being 1·01 (SD = 0·30). The agrammatic aphasic comparison group mean was 1·48 (SD = 0·80), indicating that, on average, these subjects produced a greater number of nouns as compared to verbs—a pattern typical of agrammatic aphasic subjects (see Saffran *et al.* 1989).

Our PPA subjects again showed two patterns of performance. Subjects 1, 3, and 4 demonstrated patterns quite different from Subject 2. Subject 1, at 2 years post-onset of symptoms, performed within the range of our non-brain-damaged subjects; however, on subsequent testing her noun : verb ratios became more like our agrammatic subjects' and declined to even lower levels (at 5 years post-symptoms she produced three nouns to every verb). Subjects 3 and 4 also showed marked difficulty with verb production. Subject 4 demonstrated a markedly elevated noun to verb ratio (4·40, indicating production of 4·4 nouns for every verb) at 5 years post-diagnosis, and while this pattern of verb impoverishment persisted, his noun:verb ratios actually improved on subsequent testings. This was due to a concomitant decrease in access to nouns as well; that is, his word-retrieval deficit, which initially affected primarily verbs, became much more severe and both nouns and verbs were affected.

Subject 2, in contrast, produced fewer nouns than verbs across all samples and his ratios did not change substantially over time. Noun:verb ratios ranged from 0·75 to 0·76 across the seven samples analysed.

Discussion

These results indicated two profiles of language decline in the non-fluent PPA subjects that we studied. While all subjects demonstrated reduced MLU and reduced complexity of verbal morphology compared to normal subjects, performance on several other variables differed. Three of the subjects, Subjects 1, 3, and 4, resembled our subjects with agrammatic Broca's aphasia resulting from vascular pathology; whereas one of our subjects, Subject 2, did not. His performance on several variables, including the proportion of grammatical sentences, correctness of verb morphology, ratios of open-to closed-class words and of nouns to verbs, remained only slightly below the range of our non-brain-damaged comparison groups' performance until approximately 9 years post-symptoms.

These data indicate that, within the subgroup of non-fluent PPA, the language-processing system may be affected in different ways. This finding is not surprising; indeed, we see heterogeneity in the ways in which the language-processing system is affected even in patients with acute focal vascular lesions. For example, within the classification of Broca's aphasia some patients do not present with agrammatism. Some may show marked word-retrieval difficulty affecting open-class as well as closed-class words, and no distinction between nouns and verbs. This word-retrieval deficit leads to non-fluent characteristics including pausing, incomplete or abandoned utterances, and reduced sentence length (Saffran *et al.* 1989). Other Broca's aphasic subjects show agrammatic patterns; however, different profiles of language disruption within these subjects have also been reported. That is, disruption in the generation of syntax for some of these individuals takes the form of verb-retrieval difficulty and associated problems of accessing argument structures obligated by the verb (Kegl 1995, Thompson *et al.*, 1994b). Others evince primary deficits accessing elements within the closed-class, including both bound and free-standing morphology, a deficit which also leads to a pattern of agrammatic sentence production (Menn and Obler 1989, Saffran *et al.* 1989, and others). One of our PPA subjects (Subject 2) showed behaviours more like the first pattern, while the others evinced a hybrid of the latter two patterns.

Subject 2 appeared to have difficulty accessing open-class words with both nouns and verbs being equally affected; however, access to closed-class elements, shown by his normal open:closed-class ratios and by the correctness of verb morphology, was relatively unaffected. Generation of syntax was only mildly affected at least until 9 years post-onset of symptoms. However, sentence length was abnormal even in early stages of the disorder, and at later stages abandoned utterances and inter-word pause time was quite apparent. This non-fluent pattern was also associated with articulatory programming deficits that were noted even in our earliest samples. In contrast, Subjects 1, 3, and 4 demonstrated difficulty accessing verbs and verb argument structure and, therefore, evinced difficulty generating the syntax. They also had reduced access to closed-class elements, as indicated by greatly elevated open: closed-class ratios and by frequent errors of verb morphology.

It could be argued, however, that perhaps the language-processing system was not differentially affected in our subjects, but instead that some other phenomenon may have been responsible for the differences that we observed. For example, one explanation for the deficit patterns seen in Subject 2 is that he developed compensatory strategies to overcome his aphasia; that is, perhaps he reduced his phrase length (MLU) and complexity in order to maintain grammaticality. However, Subjects 1, 3, and 4 also demonstrated reduced length and complexity of utterances, but appeared to gain no benefit in terms of sentence grammaticality. While adaptation has been proposed as one theory to explain the language deficits seen in subjects with aphasia (e.g. Kolk *et al.* 1985), the theory does not predict clearly which subjects will benefit from this strategy over others. It seems more defensible to argue that reduced MLU is an unavoidable consequence of both word-retrieval and grammatical impairments.

It also is possible that the patterns of language decline observed in our subjects are an artifact of a motor speech disorder such as dysarthria. For example, phenomena such as reduced length of utterance and production of closed-class elements may coincide with severity of dysarthria—once again, perhaps as a compensatory strategy to reduce the motoric demands of speech. We do not believe that this explanation is satisfactory for our data. Only Subject 4 presented with dysarthria at any point in the course of his progressive aphasia. Therefore, the distinction between the two profiles seen here is not supported by the presence or absence of motoric disturbance. Indeed, numbers of cases of PPA with concomitant dysarthria have been reported in the literature (Kempler *et al.* 1990, McNeil *et al.* 1995, Northern *et al.* 1990, Scully *et al.* 1986). In none of these cases was the aphasia obscured by the dysarthria.

It also could be argued that Subject 2's language patterns may not represent a different profile, but rather a slower progression of the same profile seen in the other subjects. Indeed, our data show that this was the case for Subjects 1 and 3. That is, Subject 3's decline essentially mirrored that of Subject 1, only at a later time post-onset of symptoms. In Subject 2's case, however, when his language did begin to decline more markedly (at 9–11 years post-symptom onset), his profile remained disparate. That is, like the other subjects, access to verb argument structure declined; however, unlike the others, he continued to use correct, albeit simple, verbal morphology, and normal open- to closed-class word and noun to verb ratios. In fact, his pattern of decline showed that his open-class word-retrieval deficit, affecting both nouns and verbs, worsened over time and led to a higher incidence of abandoned utterances. All the while, however, his grammatical system remained relatively spared.

Regardless of whether Subject 2's pattern of decline represents a subtype of non-fluent PPA or is merely a milder case of agrammatic-like PPA, different management approaches might have been used in his case as compared to that for Subjects 1, 3, and 4. Because Subject 2 maintained access to the grammar in the face of distinct word-retrieval deficits, this subject might have benefited—at least in the earlier years post-symptoms—from treatment focused on strategies to enhance word access. Once words were retrieved, he retained access to the grammar necessary to support sentence production. In addition, in later years he may have benefited from an augmentative communication system using a non-verbal symbol system. In contrast, Subjects 1, 2, and 3 would perhaps not have benefited from treatment focused on word retrieval; because of their grammatical involvement, simple retrieval of words might not have enhanced verbal productions. Also, in later stages of the deficit, problems with access to the grammar would preclude the use of more than a very rudimentary augmentative–alternative communication system. Of course, all these patients might have benefited from functional approaches to treatment.

The extent to which the different patterns of language decline seen in our subjects can be attributed to areas of the brain affected by the neuropathology, or to differing disease processes, however, is unknown. Studies of metabolic activity in the brains of two of our PPA subjects (Subjects 1 and 2) showed left perisylvian disturbance; whereas imaging data of one of the subjects (Subject 3) showed bifrontal abnormalities. Further, because detailed and conclusive information regarding the areas of the brain affected is available only upon autopsy, the similarities and differences of our subjects are at present unknown. The pathology responsible for the aphasias seen in our subjects also is unknown, and although it is possible that the different profiles seen in our subjects may coincide with different disease processes, this is unlikely. That is, even patients presenting with similar non-fluent PPA patterns show different neuropathology at autopsy. Of 24 non-fluent PPA subjects studied by Turner *et al.* (1996), 13 showed 'dementia lacking in distinctive histology' (DLDH) characterized by frontal lobe neuronal loss, gliosis and/or spongiform changes not associated with Alzheimer's or Pick's disease. Of the others, two had Alzheimer's disease, seven had Pick's disease or a variant, and two had questionable Creutzfeld–Jacob's disease. Indeed, further studies detailing the language decline in subjects with PPA and subsequent analysis of these subjects' brain pathology at necropsy will help us to understand the relation between site and type of neuropathology and language pathology.

Conclusions

The data reported here indicate that at least two patterns of language decline may be seen within the subgroup of non-fluent primary progressive aphasia. The first pattern resembles in many respects the deficit seen in cases of agrammatic aphasia–impaired retrieval of verbs as well as closed-class elements with consequent disruption of the sentential structures governed by those items. The second pattern seems best characterized by advancing word-retrieval difficulties with relatively little involvement of the grammatical system.

Because of the retrospective nature of this study, however, additional research is needed in order to substantiate the patterns that we observed here in other individuals with PPA. Further, additional prospective studies are needed in which data are collected under the same discourse conditions across subjects, and in which additional constrained elicitation conditions are presented. That is, even though it has been shown that the grammatical profiles of agrammatic aphasic subjects with focal lesions are similar regardless of discourse condition, discourse tasks do not provide obligatory contexts for production. Future studies comparing the language profiles of non-fluent PPA subjects at various stages in the disease process to agrammatic aphasic individuals presenting with varying degrees of severity and to other types of aphasia would also be informative.

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