Rare Lexical Speech Automatisms in a Case of Progressive Nonfluent Aphasia

Introduction

Lexical speech automatisms (LSAs) are common in aphasia following stroke (Alaquanine, 1957; Code, 1994). Extensive study in recent years demonstrates that language and speech can be impaired by progressive damage in the absence of significant impairments to other cognitive processes (Snowden, Goulding & Neary, 1989; Croot, Paterson & Hodges, 1998; Garrard & Hodges, 1999; Mesulam, 1982), but an extensive literature search has not produced a single report of speech automatisms in primary progressive aphasia (PPA). This presentation reports the analysis of the speech automatisms of a man with a progressive speech production impairment and nonfluent progressive aphasia of 10 years duration. Study of the decline of language and speech in progressive conditions promises new insights into the relationships between cognitive systems and their neural representation.

C.S. was a 62-year-old (DOB: 23/08/40), right-handed man at the time of testing. He obtained a First Class degree in chemistry and was Head of Chemistry in a well-known British public (i.e. 'private') school for 21 years before early retirement because of increasing speech problems, which were first noticed by his family in 1992. At this time too he experienced reduced sensation in parts of his upper and lower limbs and head and his family noted on a number of occasions some clouding of consciousness and verbal nonfluency on waking from a nap. Neurological investigation in 1995 included neuropsychological screening, CT scan, EEG, EMG and nerve conduction examinations that were all normal. Because there were no clear neurological reasons for his speech difficulties, C.S. was referred for psychiatric assessment that suggested anxiety and some depression. An MRI scan in March, 2001 revealed some possible mild non-localised atrophy that was considered near normal for his age and EEG was normal. Scanning in October, 2003 showed significant generalised and bilateral atrophy, more prominent in the left frontotemporal area, with indication of increased atrophy in left frontal dorsomedial regions, suggesting a predominantly fronto-temporal degeneration.

By August, 2002 a wide range of longitudinal testing had been completed, including intelligence, perception, memory, language and action/gesture. Table 1 presents a summary of cognitive assessments over the period November 2001 - August 2002. The investigations reported here took place over November 2001-February, 2002. Speech deteriorated rapidly after this, and he was virtually mute by early 2003.

We report analysis of the speech automatisms (LSAs) of C.S., with a progressive speech production impairment and nonfluent progressive aphasia of 10 years duration in the absence of significant motor or sensory impairment. This study covers the final 12 months up until he became mute. Before this time impairments of naming had emerged, but no further significant impairment to the language system.

Method & Results

At February 2002 C.S. had intact phonological, grammatical and semantic systems (agrammatic agraphia and sentence processing impairments were not noted until some months later), apart from a problem with nonword processing and naming impairments, which was more long standing. LTM, STM, working memory and perceptual processing were unimpaired and IQ was good, although may well be underestimated by the verbally dependent standardised IQ tests. C.S. had a generalised apraxia effecting limb, eye and buccofacial actions. Perception, memory and intelligence were intact. Agrammatic agraphia and sentence processing impairments began to emerge as C.S. became mute (reference withheld).

There was indication of executive impairment with disinhibition, and early indications of naming impairments, which were reported early by family members and were of more long standing. Some of C.S.'s 'anomia' may be due to speech articulation problems not recognised before this investigation began. He produced an unusual form of LSA *yes, yes* with an occasional *right*, that occurred in spontaneous speech and while responding to experimental stimuli, and this presentation reports investigations of its occurrence across tasks.

C.S. has a generalized atrophy most prominent in left frontal regions. The neural representation of speech planning and programming are still hotly debated but there is agreement that a network of functional systems are represented particularly in premotor association areas of the left frontal lobe. The medial premotor cortex of the supplementary motor area (SMA) seem to be part of an *intrinsic* system specialized for self-generated activity whereas the ventral premotor cortex (VPM), is seen as part of an *extrinsic* system specialized for motor responses to externally triggered stimulation (Jahanshahi & Frith, 1998).

C.S. LSA was first noticed by his family in 1993, so had emerged in his speech 8 to 9 years before this investigation began. It seemed to them to occur especially during more demanding speech contexts, like when C.S. when using the phone. At the time of this investigation the automatism would sometimes occur as only a single *yes*, and less frequently the word *right* and occasionally together as *yes right*. These utterances were mainly tagged onto the end of a response in single word tasks. Thus, C.S. would often respond, although with delay and hesitation, with a single word response and then tag *yes[yes/right]* on the end of this.

This presentation reports the results of an examination of LSAs in C.S.'s speech during the completion of a range of tasks increasing in length and complexity. Speech rate in speakers with nonfluent progressive aphasia have been shown to be significantly slower as tasks involve connected speech or longer speech utterances (Graham et al., 2004). This summary reports only data on picture description (picture description), connected reading (the Grandfather Passage), automatic counting (1-30), reading and repetition of high frequency single syllable words and nonwords. These tasks involved single word production and connected speech (picture description, reading a passage, counting) and they also contrast in terms of the amount of internal and external generation is entailed in their completion, allowing an examination of LSA production during an internally generated task (picture description) compared to responses to external triggering - all the other tasks are completed using different levels of external triggering. Automatic counting involves a serial recitation of probably the most over-learnt sequence of words (Lum & Ellis, 1994) stored in long-term memory. Additional longitudinal data will be analyzed subsequently.

Results of the analysis are shown in Table 2 and Figure 1. Of the 268 words produced across these tasks, 45 (16.8%) were followed by an occurrence of the automatism. There were proportionally less occurrences of the automatism in the connected reading passage (4.4%) than the single word reading and repetition tasks, (averaged across tasks at 23%) and the picture description (24%). The occurrence of the automatism in automatic counting followed the reading passage (14.3%). In the reading passage, all automatisms occurred following words with falling intonation and at this point C.S. ran out of breath. These occurrences were followed by marked delay before production of the next word.

Discussion

Why should large differences result in the number of speech automatisms that occur across different tasks? A range factors appear to account for these results. The internal generation of speech (picture description) produced more automatisms than externally triggered connected speech (reading passage and counting), but automatism production during picture description was no worse than for externally triggered single word reading and repetition, suggesting that other factors effected the emergence of automatisms. C.S.'s wpm is lowest for picture description compared to reading and counting confirming its difficulty as a task, but automatisms were no more common that in single word production, suggesting that internal and external generation were equally susceptible to automatism production. Increased complexity does not appear to influence automatism production either, as automatism production was more marked during single word reading/repetition than connected reading, a 'harder' task.

Most automatisms occurred after C.S. had successfully produced a word, and may have originally started to permeate C.S.'s speech as a confirmation of success for him, but disinhibition appears to be a likely contributor to the current results. Indeed, disinhibition has been proffered as a contributing explanation for speech automatisms since Hughlings Jackson (1879; Code, 1994). The single word tasks produced more automatisms, although appearing to entail maximum external processing, maybe because of the opportunity provided by the space at the end of the response to produce an automatism. C.S. is less able to inhibit an automatism at this time whereas during continuous connected speech C.S.'s attention is on planning and producing the next item in the string and significantly fatigued resulted from the struggle to produce utterances in a sequence. Although the picture description entails internal generation, the speech initiation and lexical access difficulties producing false starts and pauses, also provide opportunities for disinhibited automatisms to intrude.

The pattern of speech automatisms produced by C.S. do not suggest that the intrinsic or extrinsic motor speech systems are separately damaged, nor that automatisms are increased as tasks become more difficult, but do suggest that increased opportunities for disinhibition may produce more automatisms. Implications for treatment based on increasing attention and reducing disinhibition will be discussed.

References

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Table 1: Summary of comprehensive cognitive testing .

Cognitive Processes	Summary of Main Test Scores
GENERAL INTELLIGENCE	WAIS III Performance IQ: 100 Perceptual Organization IQ: 111 Working Memory Index IQ: 108 Processing Speed Index IQ: 88 Mini-Mental State Test 28/30 correct
EXECUTIVE FUNCTION	:Trailmaking Test A: 90 Secs (Scaled Score: 2) Test B: 160 Secs (Scaled Score: 4) Rey-Osterreith Complex Figure: Copy: 36/36 Short delay (3 mins): 26/36 Long delay (30 mins): 18/36 WCST - Categories Correct: 6 (Norm: 5.4) Errors: 17 (Norm: 25) Test of Everday Attention Map Search A -13 (5th Percentile)2, 28 (5th Percentile) Elevator Task - 6/7 correct Elevator Task With Distraction - 4/10
VISUAL PROCESSING	VOSP Shape Detection Test - 20/20 correct Incomplete Letters - 20/20 correct Position Discrimination - 20/20 correct Benton - Face Matching Test 20/27 correct (Long Form: 41 Normal) Ravens (Advanced) - 10/12 correct BORB Objects Tests, No Problems
MEMORY Short-term memory	WMS - III: Digit Span ñ Forward 7; Backward 3 Spatial Span: 14 SS: 10 Letter Number Sequences: 10 SS: 11 (Normal Working Memory) PALPA - Digit Matching Span - 7
Long-term memory	Hopkins Verbal Learning (Auditory presentation - Spoken responses) 10/12 correct (Written presentation/ responses) 12/12 correct Recognition: True Positives 12/12 False Positives 0/12 WMS - III:Faces I - Raw Score: 41 SS: 14 Faces II - Raw Score: 42 SS: 16 (Retention = 100%) Verbal Paired Associates I: Raw Score: 20 SS: 12 Visual Reproduction I (Recall): Raw Score: 95 SS: 14 Percentage Retention - 64% SS: 11 Recognition - 44/48 SS: 12 Visual Reproduction (Copy): Raw Score: 102 SS: 16 Historical Event Decade Judgement - 27/32 correct Warrington Face Recognition, 41/50 correct (Norm: 43/50) Warrington Word Recognition Test - 36/50 correct (Norm: 43/50)
PRAXIS	Comprehensive testing reveals significant ideomotor limb, orofacial & oculomotor apraxia with significant problems on gesture and pantomime generation to Command & to Imitation with retained gesture/pantomime comprehension and action semantics and tool use.

LANGUAGE Boston Severity Scale Fluency:	 2-3 Animals = 10 responses (Norm: 16) F.A.S. = 12 responses (Norm: 36) Proper Name Fluency - Boy's names: 9 responses Girl's names: 8 responses; Surnames: 5 responses
Semantics:	6 semantic category generation - 37 responses overall JT - Synonym Judgement Test - (Auditory) 157/160 correct, (Written) 153/160 correct No Problems on PALPA 47-49 Camel & Cactus Test - 59/64 - Shallice Synonym matching test - Easy 30/30 Difficult 25/30 Warrington matching test - Concrete 23/25, Abstract 22/25 - TOKEN (Auditory) - 31/36 correct TOKEN (Written) - 36/36 correct Synonym Judgement (Newton & Barry) 80/80 correct Proper Noun Lexical/Semantic processing
Grammatical/Syntactic	TROG 0 78/80 correct ñ 1 Block failed
Naming	Graded Naming. 17/30 correct (Average Normal) Howard Picture Naming - 40/42 correct Nickels Test, 123/130 correct McKenna Picture naming - 66/120 (Normal: 87.4 1s.d., 13.8) Hodges & Ward Familiarity Battery Faces - 30/32 (Normal: 29.3, 1s.d., 3.08) N=31 Names - 32/32 (Normal: 31.82, 1s.d., .55) N=31 Face-Name Matching, Faces - 20/22, Names - 22/22
Orthographic Processing	Lexical decision - PALPA 24 - 27 No Problems
Reading	PALPA 29 - 35 No Problems PALPA Nonword reading - 15/24 correct (Errors: 3 letters 6/6 4 letters 3/6 5 letters 4/6 6 letters 2/6) PALPA 8 Nonwords - 26/30 correct (Errors: 1 Syllable 10/10 2 Syllables 8/10 3 Syllables 8/10) NART - 30/50 correct (Full scale IQ: 111)
Repetition	PALPA 7-8 Syllable Length & Nonword, No Problems PALPA 10 - Grammatical Class, 60/60 correct; Nonword repetition, 65/80 correct' Howard syllable length, 41/42 Repetition Imag. Vs. Frequency , 77/80 (18/20 LI/LF 19/20 LI/HF) GNT Repetition test, 29/30 correct
Graphemic processing Writing to Dictation	PALPA 39 - 44, No Problems PALPA 45 Nonwords, 22/24 correct McKenna Items - 118/120 correct

Table 2. Occurrence of LSAs in different speech tasks. Wpm=words per minute.

	CS wpm	Control Means wpm	Total LSAs	Total Words & LSAs	LSAs % of total
Picture Description (19 words)	12.7	137.4	6	25	24
Reading Passage (130 words)	43.0	171.4	6	136	4.4
Auto Counting 1-30 (30 words)	26.4	105.9	5	35	14.3
Word Reading (35 words)			12	47	25.5
Word Rep (35 words)	Mean word length .478	Mean word length .623*	10	45	22.2
Nonword Rep (19 words)			6	25	24.0

- t = 6.789; p<.0001
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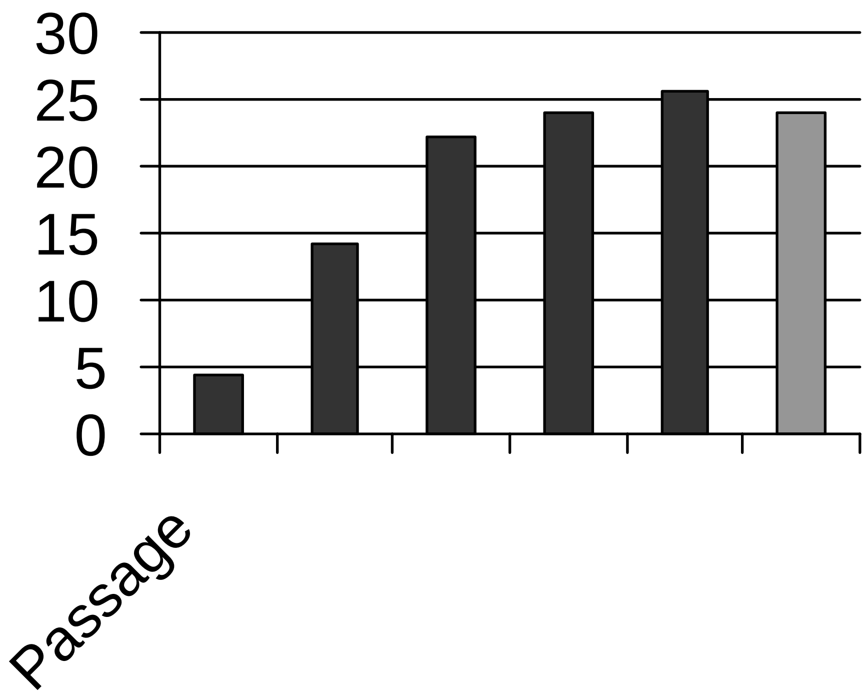

Fig 1. The percentage of total words that were speech automatisms occurring in different speech tasks. Picture description is shown in grey to highlight that it requires internal generation.