Progressive supranuclear palsy presenting with dynamic aphasia

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Abstract

Background—Progressive supranuclear palsy (PSP) is an akinetic-rigid syndrome of unknown aetiology which usually presents with a combination of unsteadiness, bradykinesia, and disordered eye movement. Speech often becomes dysarthric but language disorders are not well recognised.

Methods—Three patients with PSP (pathologically confirmed in two) were reported in which the presenting symptoms were those of difficulty with language output.

Results—Neuropsychological testing showed considerable impairment on a range of single word tasks which require active initiation and search strategies (letter and category fluency, sentence completion), and on tests of narrative language production. By contrast, naming from pictures and from verbal descriptions, and word and sentence comprehension were largely intact. The degree of semantic memory impairment was also slight.

Conclusions—Relatively selective involvement of cognitive processes critical for planning and initiating language output may occur in some patients with PSP. This presentation resembles the phenomenon of “verbal adynamia” or “dynamic aphasia” seen in patients with frontal lobe damage. Although definite cortical changes were present at postmortem examination, it is likely that the neuropsychological deficits reflect functional frontal deafferentation secondary to interruption of frontostriatal feedback loops.

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Keywords: progressive supranuclear palsy; dynamic aphasia

Progressive supranuclear palsy (PSP) as originally described by Richardson et al in the 1960s characteristically presents as a parkinsonian syndrome with axial rigidity, bradykinesia, postural instability, and paralysis of conjugate gaze for voluntarily guided eye movements. Dysarthria and swallowing difficulties occur as a result of involvement of the muscles of articulation by extrapyramidal rigidity. Although dysphasia (of a generally unspecified type) and word finding difficulties have been noted in some patients with PSP during the course of their illness,2,3 descriptions of a presentation with a disorder of spoken language production are virtually absent from the medical literature; Perkin et al reported five patients with atypical presentations, two of whom had severe dysphasia in the context of mild global dementia, one made naming errors and later developed unintelligible speech, and the other was described as a non-fluent dysphasic. The lack of awareness of this aspect is illustrated by the fact that two recent books devoted entirely to PSP,4,5 although mentioning mild word finding difficulty and reduction in speech output during the course of the disease, do not refer to this presenting feature. We have seen three patients with PSP in whom the initial presentation was one of a verbal adynamia, resembling the phenomenon described by Luria,6 also sometimes termed dynamic aphasia.7 All three underwent detailed neuropsychological and linguistic assessment, and in two the diagnosis was confirmed pathologically.

Patient 1

A 62 year old gardener presented in 1990 to a psychiatrist after a two year history of a change in personality. Initially, he had been disinhibited and aggressive, but had subsequently become anergic and apathetic. Over this period it was noticed by the patient, and his family, that his expressive output in both speech and writing had diminished greatly. Practical abilities around the home seemed preserved. Examination in 1991 showed him to be very non-fluent with pronounced word finding difficulties. Speech was limited to single words or short phrases which were grammatically correct without phonological or semantic paraphasias. By contrast, confrontation naming, word and sentence repetition, and comprehension of simple and complex commands were strikingly normal. Physical examination showed a mild degree of supranuclear vertical gaze limitation and poor horizontal saccadic movements. His gait was wide based and slightly unsteady, but Romberg’s test was negative. Other extrapyramidal signs were absent.

Brain CT showed a mild degree of frontal atrophy. Examination of CSF was normal. The diagnosis at this stage was unclear, but PSP or dementia of frontal lobe type were considered the most likely possibilities.

Over the ensuing 12 months, anxiety and a preoccupation with death became prominent
features. Axial rigidity became pronounced, and there was a loss of all voluntary eye movements. He became increasingly immobile and often fell. Speech was restricted to single word repetitive utterances before mutism supervened in the terminal stages of his illness. He died four years after the onset of the first symptoms.

POSTMORTEM BRAIN EXAMINATION
Macroscopical examination of the brain showed a degree of cerebral gyral atrophy, more pronounced in the lateral frontal regions, and pallor of the substantia nigra and locus coeruleus in the brain stem. Microscopically, severe nerve cell loss and gliosis were evident in the subthalamic nucleus and the central mesencephalic grey. Less severe neuronal loss was present in the brain stem, pigmented nuclei, and the dentate nucleus. Surviving nerve cells in these subcortical and brain stem areas contained tau positive neurofibrillary tangles; tangles were also present in the globus pallidus, the red nucleus, the brain stem reticular formation, the pontine nuclei, and particularly in the nucleus of Darkshevich in the rostral midbrain. Neurofibrillary tangles were numerous in the frontal cortex but there were very few in the temporal cortex; senile plaques were sparse. The type and distribution of lesions confirmed the diagnosis of PSP.

Patient 2
A retired dispatch manager presented at the age of 67 when his wife noticed that over the previous two years there had been a reduction in his speech, which was now reduced to single words or short phrases. His comprehension seemed intact and he was only mildly forgetful. On further enquiry, it became apparent that more recently his personality had changed to the extent that he had become rigid, irritable, and had lost his sense of humour, but he did not complain of any of these difficulties spontaneously. His general abilities around the home seemed normal, and he had been able to cook and clean when left on his own while his wife was in hospital for a cataract operation. A degree of postural instability was evident from the fact that he had had several falls.

He had a dyspraxic gait and postural instability. Eye movements were abnormal with reduced down gaze pursuit and very slow voluntarily guided horizontal and vertical saccades. He had prominent pout and bilateral grasp reflexes. His speech was appropriate but very reduced in output, consisting of short, grammatically correct phrases. Paraphasic errors were absent. Naming, word and sentence repetition, and comprehension were normal. He was fully oriented and showed good recall of novel verbal material (a fictional name and address).

Brain CT showed mild frontal lobe atrophy, and CSF examination was normal.

Over the subsequent three years, his condition steadily deteriorated, and there was no response to treatment with levodopa. He became dysphagic, mute, and immobile and died after a total duration of illness of five years.

POSTMORTEM EXAMINATION
Macroscopically, the brain showed mild generalised gyral atrophy and pallor of the substantia nigra. Microscopical examination showed a few neurofibrillary tangles in the frontal cortex and an occasional tangle in the temporal cortex. Senile plaques were absent. Nerve cell loss and gliosis were present in the subthalamic nucleus. Neurofibrillary tangles were numerous in the globus pallidus, Meynert’s nucleus, the thalamus, the hypothalamus, and the subthalamic nucleus. Tangles were also prominent in the red nucleus, substantia nigra, the nucleus of Darksheivich, central mesencephalic grey, brain stem pigmented nuclei, reticular formation, pontine nuclei, accessory dorsal olivary nucleus, and the dentate nucleus. As in the previous patient, the neuropathological findings were diagnostic of PSP.

Patient 3
A right handed housewife and exwarden of a nursing home presented at the age of 65 with a one year history of word finding difficulty in speech and very impaired conversational abilities. Her ability to use her right hand in writing had slowed up and there was a tendency to overbalance easily. Despite the reduced verbal output and clumsiness, she continued to complete her daily crossword puzzle.

She had somewhat immobile facies, mild axial rigidity, and a hesitant gait. Voluntary upward saccades and pursuit movements were abnormal. Her speech was mildly dysarthric and slow, with reduced phrase length. Paraphasic errors were absent. Naming, word and sentence repetition, and comprehension were normal.

Brain CT, chest radiograph, thyroid function, syphilis serology, and CSF examination were all normal.

She was treated with levodopa, lisuride, and amantadine, without noticeable response and her condition deteriorated. Five years from the onset of first symptoms, she was chairbound with very dysarthric speech and minimal spontaneous utterances. Rigidity was pronounced. Occasional instances of inability to inhibit an initiated movement were seen, for example, repetitive hand rubbing. Voluntary conjugate eye movements were severely restricted, especially in the vertical plane.

On the basis of the criteria used by Golbe et al., she conformed to a clinical diagnosis of PSP (supranuclear gaze palsy, onset after 40, progressive course, bradykinesia, rigidity greater axially than in the limbs, dysarthria, and the absence of tremor).

Neuropsychological assessment
A battery of neuropsychological tests was given to each patient to assess a broad range of cognitive abilities including memory, visuo-perceptual functions, expressive language,
comprehension, and semantic knowledge.

The patients' results are illustrated in the accompanying tables and are compared with a group of 24 normal volunteer subjects (18 women, six men) with mean age 69-7 (SD 7-8) years, and mean 10-8 (SD 2-3) years education selected from the MRC Applied Psychology Unit's subject panel.

GENERAL COGNITION
Assessment of global cognitive function was restricted in patient 1, and to a lesser extent in patient 2, by their severe communication difficulties. Patient 3 performed normally on the mini mental state examination\textsuperscript{14} and the dementia rating scale,\textsuperscript{15} whereas patient 2 showed a mild degree of impairment on both measures. All three showed a significant decline on these measures late in the course of the disease (table 1).

MEMORY
Immediate auditory-verbal short-term memory, as measured by digit span forwards, was normal at presentation in all three patients, but became impaired as their disease progressed (table 1). Tests of longer term verbal and visual memory showed more pronounced impairment, based on both recall (logical memory)\textsuperscript{16} and recognition paradigms,\textsuperscript{17} in two of the three patients. Interpretation of the logical memory results was, however, complicated by the patients' spoken language deficits. Although ability to copy the Rey figure\textsuperscript{18} was impaired, it is notable that the percentage recalled was similar to that of controls.

PERCEPTION
Basic perceptual abilities were assessed with an object matching task\textsuperscript{19} in which the subject is presented simultaneously with three pictures consisting of a photograph of a common object together with a photograph of the same object taken from a different angle and a visually similar foil. Patients 2 and 3 performed normally at presentation but the performance of patient 2 subsequently declined on follow up.

Table 1: General cognition and memory

<table>
<thead>
<tr>
<th>Duration of disease (months)</th>
<th>MMSE</th>
<th>Dementia rating scale</th>
<th>Logical memory (immediate)</th>
<th>Logical memory (delayed)</th>
<th>Recognition memory (words)</th>
<th>Recognition memory (faces)</th>
<th>Digits forward</th>
<th>Digits backward</th>
<th>Rey copy</th>
<th>Rey recall</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 1</td>
<td>36</td>
<td>22</td>
<td>5-75</td>
<td>2-25</td>
<td>33</td>
<td>28</td>
<td>6</td>
<td>4</td>
<td>2-5</td>
<td>NT</td>
</tr>
<tr>
<td>Patient 2</td>
<td>24</td>
<td>24</td>
<td>12-4</td>
<td>1-5</td>
<td>26</td>
<td>28</td>
<td>6</td>
<td>4</td>
<td>11</td>
<td>NT</td>
</tr>
<tr>
<td>Patient 3</td>
<td>48</td>
<td>21</td>
<td>8-0</td>
<td>2-25</td>
<td>NT</td>
<td>NT</td>
<td>6</td>
<td>2</td>
<td>NT</td>
<td>NT</td>
</tr>
<tr>
<td>Maximum score</td>
<td>30</td>
<td>101</td>
<td>3-75</td>
<td>1-5</td>
<td>NT</td>
<td>NT</td>
<td>NT</td>
<td>NT</td>
<td>NT</td>
<td>NT</td>
</tr>
<tr>
<td>Controls</td>
<td>29-2</td>
<td>144</td>
<td>23-2</td>
<td>17</td>
<td>43-7</td>
<td>6-8</td>
<td>48</td>
<td>34</td>
<td>15-5</td>
<td>14</td>
</tr>
</tbody>
</table>

Table 2: Language: single-word production tasks

<table>
<thead>
<tr>
<th>Duration of disease (months)</th>
<th>Letter fluency</th>
<th>Category fluency</th>
<th>Naming pictures</th>
<th>Naming description</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Letter fluency</td>
<td>Living × 4</td>
<td>Mammade × 4</td>
<td></td>
</tr>
<tr>
<td>Patient 1</td>
<td>36</td>
<td>3</td>
<td>15</td>
<td>36</td>
</tr>
<tr>
<td>Patient 2</td>
<td>30</td>
<td>6</td>
<td>27</td>
<td>20</td>
</tr>
<tr>
<td>Patient 3</td>
<td>18</td>
<td>27</td>
<td>23</td>
<td>45</td>
</tr>
<tr>
<td>Maximum score</td>
<td>28</td>
<td>10</td>
<td>23</td>
<td>24</td>
</tr>
</tbody>
</table>

NT = Not tested;
MMSE = mini mental state examination.

Expressive language abilities
SINGLE-WORD PRODUCTION TASKS: VERBAL FLUENCY AND NAMING ABILITIES
One of the most striking dissociations in all three patients was that between confrontation naming and verbal fluency. Two commonly used verbal fluency tasks—letter and category fluency—were given to all three patients. For letter fluency, they were asked to generate as many words as possible beginning with the letters "F, A, and S" excluding proper nouns and the same word with different suffixes (fix, fixed, fixing, etc). For category fluency tests, subjects were given one minute for each of four "living" categories (animals, birds, water creatures, and breeds of dog) and four "manmade" categories (household objects, vehicles, musical instruments, and types of boat). On both initial letter and semantic category based tasks all three patients showed very pronounced impairment with a progressive decline over time. The normal relation of better performance on category fluency was maintained in all three (table 2).

The drastically impaired verbal fluency contrasted with their normal, or near normal, performance on tests of naming ability, using as
stimuli either 48 line drawings of animals and objects or 24 verbal descriptions (for example, “What do we call a small vehicle with runners, used on snow or ice”—answer: sledge); the verbal description tasks were taken from the semantic battery which has previously been described in detail. Examination of the errors made on picture naming showed that most were either visual confusions (for example, “a row of books” for accordion) or circumlocutions (“shellfish with claws” for lobster).

**Narrative and Discourse Abilities**

As one of the principal complaints of the patients’ families was the patients’ lack of conversation, we applied some measures aimed at assessing narrative language skills.

The cookie theft picture description task requires subjects to produce a verbal description of a drawing of a domestic scene. All three patients showed considerable poverty of language output when presented with this task and also tended to perseverate. Their responses were analysed according to the number of T units and morphemes they contained. A T unit is an independent clause (a verb and a noun) with its dependent clauses. A morpheme is the smallest unit of meaning (for example, “house” = 1 morpheme, “houses” = 2 morphemes, “give” = 1 morpheme, “gave” = 2 morphemes). Transcriptions of the patients’ descriptions are reproduced in the appendix with the T unit and morpheme scores.

Patients 1 and 3 showed a progressive decline in the total number of morphemes produced, but in patient 3 the absolute number of T units remained fairly constant over this period resulting in a steady fall in the ratio of morphemes to T units. Other features of the patients’ narratives are the occasional syntactic errors with omission of appropriate function words (particularly hiding in patient 2), the pauses while searching for words resulting in aborted phrases, and the tendency to perseveration.

To assess narrative language skills in a more naturalistic setting, we applied the Amsterdam-Nijmegen everyday language test, which has been extensively used to monitor recovery in patients with aphasic stroke. In this task, the subject is asked to describe in 10 everyday situations, which they then describe in appropriate conversational speech to the examiner. For example:

1. Examiner: “You have just moved to my street, and you would like to meet me. You ring my doorbell and say?"
   Patient 2: “Meet me. Meet me. Meet me.”
   Patient 3: “How about meeting for a cuppa?”

2. Examiner: “You see your neighbour walking by. You want to ask him to visit you sometime. What do you say?”
   Patient 2: “Visit me. Visit me . . . That’s all”
   Patient 3: “Could you come to visit me sometime?”

3. Examiner: “You are at the butcher’s and this is lying on the floor (a glove). What do you say?”
   Patient 2: “That’s not mine.”
   Patient 3: “Somebody left a glove . . . a sheepskin glove . . . somebody has dropped it.”

Each of the 10 responses were scored for understandability (information content and ability to convey the appropriate message) and intelligibility (the clarity of speech in terms of phonetic, articulatory and prosodic components) on a five point scale (total score for understandability = 50) according to the criteria applied by Blomert et al. Controls obtained mean scores of 95·6 (SD 4·8)% and 91·6 (SD 11·5)% for understandability and intelligibility respectively. The content of the responses of patient 2 was reduced, and although appropriate, lacked normal conversational richness with a resultant laconic and sometimes telegraphic style; he scored 62% on both understandability and intelligibility of the message. On a second administration of the test, eight months later, perseveration was prominent, with a tendency to echo the words used by the examiner; his scores at this stage had fallen to 40% and 20%. Patient 3 scored within the normal range (86% for understandability and 100% for intelligibility). Patient 1 was not available for testing.

**Sentence Completion Tasks**

To evaluate word finding and production ability in a task which requires a constrained, but untimed semantic search, we used a sentence completion test based on that used by Costello and Warrington in their examination of a patient with dynamic aphasia resulting from a left frontal astrocytoma. In this task, the subjects were given the stem of a sentence and then asked to provide an appropriate single word to complete it. Twenty sentences required a noun for completion, and 20 a verb. In a half of the noun and verb completion sentences, the possible choice was limited by the stem (closed sentences—for example, “Most cats see very well at . . . ”, “He mailed the letter without a . . . ”), and in the remainder, the choice was wide (open sentences—for example, “The man with a moustache wore a . . . ”, “Clumsily the woman loosened her . . . ”). The control subjects had no difficulty with this task, and produced an average of only 1·1 (2·7%) errors for the 40 sentences which consisted of violations of the task requirements by adding more than one word to complete the sentence, or occasional failures to produce a response. Patient 2 made 12 errors (30%), tending to perseverate words from the sentence stem, or the previous response. Patient 3 also made 12 (30%) errors, violating the rules of the task consistently by producing short phrases (rather than words). Patient 1 was unavailable by the time we devised this task.

**Phrase Completions**

In a variant of the above task, we asked subjects to complete 40 sentences with a short phrase. Controls made an average of 0·6 (1·5%) errors which were either violations or
non-responses. Patient 2 produced nine bizarre responses (22.5%) which failed to take account of the prosodic and semantic information in the cue (for example, “The girls had tried hard but”... “freedom”; “The business had crumbled so”... “the boy thought”; “The husband disappeared last year and”... “a war”). Patient 3 was unable to formulate a response in 12 out of the 40 (30%) sentences, but did not produce any erroneous responses.

**Semantic and syntactic comprehension**

Comprehension of single word meaning was largely intact in all three patients. On the word picture matching test from the psycholinguistic assessments of language processing in aphasia,26 in which each target item is displayed together with a close and distant semantic foil, a visually related foil, and an unrelated foil, two of the patients achieved near perfect scores (table 3). Likewise, on the word-picture matching subtest from the semantic battery27 28 in which subjects are presented with eight target line drawings that belong to the same category on an A4 sheet and asked to point to one named by the examiner (for example, eight different animals: deer, rabbit, horse, cow, squirrel, fox, cat, and mouse), the same two patients (2 and 3) performed normally. The impaired performance of patient 1, and the slight subsequent decline in patient 2, may reflect their considerable problems with visual scanning.

Conceptual or semantic knowledge was probed with other subtests of the semantic battery, which assesses knowledge about the same 48 items—half living and half manufactured—across a range of tests.21 22 In the sorting task, the subject has to sort picture cards into defined categories at three levels (level 1: living v manmade; level 2: land animal v bird v water creature; household item v vehicle v musical instrument; level 3: native v foreign animal, fierce v non-fierce animal, electrical v non-electrical item, etc). All three performed normally on this task at presentation, but in two patients (1 and 2), performance declined as their disease progressed.

In the semantic feature questionnaire from the same battery, the subject is asked four questions relating to physical characteristics (for example, “Is an elephant grey?”) and four relating to functional attributes (for example, “Does an elephant eat meat?”) of 24 items used in the naming test. Four of the questions are designed to elicit a “yes” answer, and four a “no” response. Two patients (2 and 3) performed normally on this task (table 3), and although patient 1 (and subsequently patient 2) produced results outside the normal range, there was evidence that they continued to retain a significant amount of semantic knowledge (77% and 80% of maximum score respectively on their worst performances).

The definitions subtest was given to each patient at presentation. In this test, subjects are asked to generate definitions of 12 of the items in the battery in response to the spoken word, each within the space of one minute. Normal subjects produce a mean of 6-8 pieces of factual information with hardly any perseverative or intrusive errors.21 The information supplied by each of the three patients was correct, but there was a diminution in the amount produced (mean of 2-6, 5-6, and 5-1 in patients 1 to 3 respectively) with frequent perseveration. For example:

**Patient 1:**

Seal: “You pull it.”


Accordion: “Buttons on it... got buttons on it, squeeze-box, squeeze it. Got it in front of me.”

**Patient 2:**

Crocodile: “An animal, it’s got skin, lovely skin, it’s got four feet, it swims, it’s got a tail. Eats man. Water—it likes water.”


**Patient 3:**

Peacock: “A bird. Has a big tail which it fans out when its mating. They’re quite big.”

Toaster: “Household item. Can be for two or four pieces of toast. Its either pop-up or manually operated.”

Comprehension of syntactic structures was tested using the test for the reception of grammar.23 In this test, the subject is asked to point to one of four pictures in response to phrases or sentences of increasing syntactic complexity in which the vocabulary remains very simple (for example, “The boys pick the apples”; “The boy is chased by the dog”; “The pencil is on the book that is yellow” etc). In total, there are 80 arrays divided into 20 blocks, each of which tests comprehension of a particular syntactic structure (for example, plurality, gender, locative prepositions, negatives, reversible

<table>
<thead>
<tr>
<th>Table 3 Semantic and syntactic comprehension</th>
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<tbody>
<tr>
<td>Duration of disease (months)</td>
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<tr>
<td>--------------------------------</td>
</tr>
<tr>
<td>Case 1</td>
</tr>
<tr>
<td>Case 2</td>
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<td>Case 3</td>
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<td>Maximum score</td>
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<td>Controls (SD)</td>
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</table>

NT = Not tested; PALPA = psycholinguistic assessments of language processing in aphasia.
active, reversible passive, postmodified subject, relative and embedded sentences). One patient (3) obtained a near perfect score but the other two showed evidence of mild impairment.

Discussion

The three patients all presented with a considerable reduction in verbal output resulting in difficulties with propositional language. With disease progression, their speech became radically impoverished with eventual muteness. At a stage when conversational language and verbal fluency were strikingly impaired they exhibited normal, or near normal, performance on tests requiring naming from pictures and from oral descriptions, as well as tests of word and sentence comprehension. Repetition of words and sentences also seemed to be intact, although this was not tested formally. Phonological speech errors never occurred. This pattern is best described as verbal adynamia or dynamic aphasia.20-22 Although dysphasia is classically regarding as a feature of cortical dementias (such as Alzheimer's disease), rather than diseases which involve predominantly subcortical structures, as noted earlier dysphasia has been reported by several investigators during the course of PSP,23-24 and on occasions has been described as a prominent feature at presentation.7 The nature of the language disturbance in these patients has, however, not been analysed in any detail and to the best of our knowledge, PSP presenting with dynamic aphasia has not been recorded previously. It should be noted that subtle changes in personality were also apparent to the relatives of the first two patients.

The reduction in language output in these patients progressed in parallel with the time course of their disease, and was greatest on tests requiring the subject to generate discourse, such as the Amsterdam-Nijmegen everyday language test,25 or narrative as in the cookie theft picture.26 Verbal output on these tasks was impoverished; the patients were able to produce appropriate information but could often manage to communicate with incomplete sentences only. Although almost all elements of the cookie theft picture were mentioned, the information produced was insufficient to describe fully the situation presented. As can be seen from the performances of patient 3 on serial administration of the picture, there was a progressive decline in her output, as evidenced by the progressive reduction in morphemes/T unit.27 28 Tasks of sentence completion also proved difficult for the two patients tested: they produced an abnormally large number of omissions and occasional bizarre responses. With disease progression, there was a pronounced tendency to perseverate using responses from previous questions, or to produce echolalic responses with repetition of the sentence stem. When the patients were required to produce more than one word in response to the stem, which might be expected to place less of a constraint on performance because there is more opportunity to vary the phrase, this difficulty persisted.

One of the other striking findings in our patients was the degree of impairment on both letter and category based verbal fluency. Normal subjects perform better on category than on letter based tests.29-32 Although clearly very poor at both tasks, the “normal” relation between the two tasks was maintained in all three patients with PSP, a finding in keeping with that of a larger group of patients with PSP.33 This contrasts with the pattern seen in Alzheimer's disease in which category fluency is consistently poorer than letter fluency.29-32 It has been argued that the deficit in letter fluency reflects the early breakdown of semantic memory found in Alzheimer's disease, whereas the equal impairment in letter and category fluency found in PSP probably results from impaired initiation and retrieval processes, which are common to both tasks.

Against this background of grossly impaired performance on tests of letter and category vocabulary, the occasional oddities in completion tasks which is a feature of cortical dementias is notable that all three patients performed within normal limits on a conventional 48 item picture naming test and on a test requiring subjects to name from verbal descriptions. On confrontation naming, any errors reflected perceptual difficulties or mild impairment in access to phonological word forms (circumlocutions). Semantic memory, as measured by performance on tasks which require subjects to access the central representational knowledge concerning a range of living and man-made items, was only mildly affected in our patients. They performed normally on two tasks requiring the matching of spoken words to pictures, although it should be pointed out that such tasks are relatively insensitive to semantic memory impairment.22 On the more demanding picture sorting and semantic feature questionnaire subtests of the semantic test battery, our patients showed a mild degree of impairment in keeping with the results of a larger scale group study of patients with PSP.33

The difficulties noted with language production cannot, therefore, be adequately explained by a loss of semantic information; it has been repeatedly shown that even a mild degree of semantic disruption causes substantial impairment on standard tests of confrontation naming. A major phonological or articulatory deficit can also be excluded, and although the patients did show abnormalities of syntactic structure on elicited speech production tasks, a primary disorder of syntax cannot explain their profound difficulties with narrative speech production, letter and category fluency, and sentence completion tasks.

The characteristics of the language abnormality in our cases most closely resembles that described by Luria34 who delineated the features of “dynamic aphasia” in patients with traumatic frontal lobe damage. Luria reported that such patients have no difficulty with naming or repetition, but find expression of internally generated thoughts and the production of simple verbal statements very difficult due to a "disturbance of the linear scheme of a sen-
Progressive supranuclear palsy presenting with dynamic aphasia

tence”. In a more recent case study, Costello and Warrington described a patient with a large left frontal tumour who had intact naming and reading skills, but an almost total lack of spontaneous speech. They argued that Luria’s concept of dynamic aphasia was inappropriate in their case, and that instead it was a failure at the “higher order” stage of initial formulation of a phrase or sentence. It would seem, therefore, that in some patients with dynamic aphasia there might be a breakdown at the highest “pragmatic” level of language planning—that is to say in converting thoughts to speech acts—whereas in others the functional locus might be at a lower level of word selection, sentence planning, and implementation.

Although slowness in speech production and dysarthria have been reported in PSP by some authors, more profound linguistic defects have been recorded only rarely. For instance, the six patients studied by Podoll et al had abnormalities of speech production that were characterised by short simple sentences and a tendency to leave sentences unfinished; but semantics, syntax, and grammar were found to be normal. The difficulties in language production were considered by the authors to be non-specific and secondary to a combination of extrapyramidal features and dysarthria. Maher and Lees enumerated the clinical features in 52 patients with PSP and noted dysphasia in four, but the nature of the language disturbance was not further elucidated. In a paper discussing brain neurotransmitter levels in five patients with pathologically established PSP, Kish et al described severe aphasia with paraphasic word substitutions developing in one patient in the last year of her life. One of the patients reported in a PET study by Leenders et al had evidence of dysphasia but no further details were provided. In a detailed single-case study of language in PSP, Lebrun et al described a 55 year old man whose symptoms included speech difficulties resembling stuttering with a degree of dysphonia and a tendency to perseverate on the last word or syllable in a phrase. Neuropsychological evaluation showed intact comprehension and syntax in spoken speech, and choice of words was appropriate. More recently, Daniele et al reported a patient with a clinical diagnosis of PSP who had presented with dysarthria and difficulty writing. More detailed assessment disclosed word finding difficulty with semantic errors and omissions on confrontation naming as well as other deficits affecting particularly verb, rather than noun, processing.

It is possible that our patients represent a subgroup of PSP in whom language is particularly affected. Alternatively, deficits in language production may be more common than is realised but may have been overlooked because narrative abilities have not been investigated.

Turning to the question of the neuroanatomical and pathological basis for the dynamic aphasia, PSP is traditionally regarded as a prototypic form of subcortical dementia. More contemporary neuropathological studies have, however, shown subtle but definite cortical changes in PSP consisting of neocortical neurofibrillary tangles of the globose type affecting the larger pyramidal and small neurons and most often involving the precentral gyrus. A very recent study of 17 patients with pathologically confirmed Steele-Richardson-Olszewski syndrome (a designation preferred by the authors since not all patients had supranuclear gaze palsies) again reported the presence of mild cortical pathology that did not seem to correlate with the degree of dementia. It is interesting to note that all of the patients with supranuclear gaze palsies became demented whereas a lesser proportion of those without ocular-motor abnormalities did so, suggesting that the cognitive deficits are indeed related more to subcortical than cortical involvement.

Both of our patients who underwent post-mortem examination had a degree of generalised cortical atrophy and showed specific neurofibrillary pathology in the subcortical cortex. Despite these cortical changes, the brunt of the neuronal loss and globose tangle formation in PSP was in subcortical structures, such as the substantia nigra, subthalamic nucleus, nucleus basalis of Meynert, globus pallidus, corpus striatum, cerebellum, and various brainstem nuclei. It seems likely, therefore, that subcortical structures were the site of initial pathology and that the dynamic aphasia in our patients is likely to reflect the interruption of frontostriatal loops rather than direct cortical pathology. Brain PET studies of patients with PSP have shown low striatal and frontal uptake of 18F-2-fluorodeoxyglucose, and reduced putamen and caudate uptake of 18F-6-fluorodopa; these findings have been interpreted as supporting the notion that the frontal dysfunction in PSP is secondary to pathology in the basal ganglia.

In summary, we have presented three cases of PSP in whom a disturbance of language output was the presenting complaint. Further analysis of their difficulties has led us to believe that there may be a failure of language production which is most saliently attributable to a semantic or phonological dysfunction or by a fundamental loss of semantic knowledge. The occurrence of bizarre responses in some of the tests may reflect a failure of executive functions responsible for overall supervision of appropriateness. This phenomenon is well recognised in other diseases with predominant pathology in the frontal lobes. The language profile seen most closely resembles dynamic aphasia, which has been shown to occur in patients with frontal lobe disease and reflects a breakdown at a high level of language planning and initiation. Dynamic aphasia may be more common in PSP than has been realised previously. Many issues remain to be considered in future studies, including the fundamental question of whether this dynamic aphasia represents a breakdown in all aspects of higher order thought, language, and action planning, or is restricted to the domain of expressive language. It also remains open to question whether the observed neuropsychological pro-
file reflects functional frontal deafferentation secondary to the interruption of frontotriatal feedback loops, or direct frontal cortical involvement, although the balance of evidence perhaps favours the first explanation.

Appendix

PATIENT 1

(1) 36 months duration of illness

"The little boy's nearly toppling over/ the cupboard's open/ the wrong way/washing up/ the wrong way/ cup/ board's open/ the wrong way/ the boy's stool/ the boy's stool started to back up." Total T units = 5. Total morphemes in T units = 36. Morphemes/T unit = 7.2.

(2) 41 months duration of illness

"Everything I see going on in the picture... yeah... yeah... (mumbles) hungry/washing up (mumbles)." Total T units = 0 (no descriptive information).

PATIENT 2

(1) 40 months duration of illness

"Washin up... overflowing... cookie jar... washing the girl's hand up, and window, and tree, and curtains/she's got shoes on... a leg crossed... water overflowing/ got the taps on/ that's wrong." Total T units = 7. Total morphemes in T units = 31. Morphemes/T unit = 4.4.

(2) 18 months duration of illness

"Well the sink is overflowing and the housewife doesn't seem to be taking much notice/she's washing up, wiping up, and the boy is stealing from the cookie jar but he's fallen over because he's on a stool/I can't think of anything else." Total T units = 5. Total morphemes = 50. Morphemes/T unit = 10.0.

(3) 28 months duration of illness

"The housewife is pinning the cookie jar... cookies from the jar and giving them to the girl and he's fallen over with the stool um... the mother is er... having a flood... the er... kitchen sink is overflowing/ the curtains aren't pulled very nicely." Total T units = 5. Total morphemes = 39. Morphemes/T unit = 6.5.

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Progressive supranuclear palsy presenting with dynamic aphasia.

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