Trying to tell a tale

Discourse impairments in progressive aphasia and frontotemporal dementia

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Abstract—*Objective:* To assess discourse in patients with frontotemporal dementia (FTD). *Methods:* The authors asked patients with progressive nonfluent aphasia (PNFA), patients with semantic dementia (SemD), and nonaphasic patients with a disorder of social comportment and executive functioning (SOC/EXEC) to narrate the story of a wordless children's picture book. *Results:* The authors found significant discourse impairments in all three groups of patients. Moreover, there were qualitatively important differences between the groups. Patients with PNFA had the sparsest output, producing narratives with the fewest words per minute. Patients with SemD had difficulty retrieving words needed to tell their narratives. Though not aphasic, patients with SOC/EXEC had profound difficulty organizing their narratives, and they could not effectively express the point of the story. This deficit correlated with poor performance on a measure of executive resources requiring an organized mental search. In addition, a correlation of narrative organization with cortical atrophy in patients with SOC/EXEC was significant in right frontal and anterior temporal brain regions. *Conclusions:* Impaired day-to-day communication in nonaphasic frontotemporal dementia patients with a disorder of social comportment and executive functioning is due in part to a striking deficit in discourse organization associated with right frontotemporal disease. Difficulty with discourse in progressive aphasia is due largely to the language impairments of these patients.

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Self-expression in speech is a critical feature defining the humanness of our species. Discourse is the construction of the overall theme or goal that organizes a sequence of utterances in a coherent manner,[1,2] and a discourse impairment devastates an individual's ability to relate to others. In this study, we assess discourse in patients with frontotemporal dementia (FTD).

Frontotemporal dementia affects behavior and language but entails little memory impairment.[3,4] Discourse has not been studied previously in FTD, even though the families of patients with aphasic and nonaphasic FTD alike frequently report conversational difficulty. Although it is not difficult to imagine that a language disturbance can interfere with narrative communication at the single-word or sentence level,[5-7] it is the right hemisphere that is compromised in FTD patients with a disorder of social comportment and executive difficulty (SOC/EXEC).[8-10] Studies of nonaphasic stroke patients with a discourse impairment also implicate this region.[11,12] Impairment of executive resources such as planning and organization that are compromised in patients with SOC/EXEC may contribute to their discourse difficulty.[13-15]

In this study, we examined narratives told by patients from a wordless picture book. We expected that patients with progressive nonfluent aphasia (PNFA) would produce fewer words, whereas patients with semantic dementia (SemD) would have difficulty due to limited lexical retrieval. SOC/EXEC patients, although not aphasic, were expected to have impaired discourse due to poor organization, possibly related to limited executive resources. We also examined the relation of impaired narrative organization in patients with SOC/EXEC to right frontotemporal disease.

Methods. *Subjects.* We studied 35 patients with FTD (10 PNFA, 13 SemD, and 12 SOC/EXEC) and 10 healthy seniors. Patients with FTD were diagnosed by an experienced neurologist (M.G.) according to published criteria.[16,17] The assignment of patients to FTD subgroups was based on a consensus of two independent raters reviewing a semistructured neurologic history, a complete neurologic examination, and a detailed mental status examination according to a revision[18-20] of the Neary et al. clinical diagnostic criteria.[21] Exclusionary criteria included other causes of dementia, such as metabolic, endocrine, vascular, structural, nu-

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Table 1 Mean (SD) demographic and clinical characteristics of patients with frontotemporal dementia and controls

	Controls	PNFA	SemD	SOC/EXEC
No. male/female	2/8	3/7	9/4	7/5
Age, y	69.1 (4.8)	72.5 (7.6)	66.4 (7.1)	64.8 (13.1)
Education, y	16.7 (2.6)	14.8 (2.9)	15.8 (2.7)	16.0 (3.0)
MMSE, max = 30	30.0 (0.0)	25.7 (3.9)	22.4 (7.8)	25.0 (6.8)
Disease duration, y	—	3.5 (1.2)	4.6 (2.1)	4.5 (1.5)
FAS performance, 3 minutes	44.8 (11.2)	13.3 (6.2)	22.1 (12.5)	18.5 (13.6)

PNFA = progressive nonfluent aphasia; SemD = semantic dementia; SOC/EXEC = disorder of social comportment and executive functioning; MMSE = Mini-Mental State Examination.

tritional, and infectious etiologies, and primary psychiatric disorders. We also excluded patients who had visual-perceptual difficulty that limited their ability to perceive the pictures. A one-way analysis of variance (ANOVA) indicated that all four subject groups were matched for age and education. The patients were mildly impaired by the standard of the Mini-Mental State Examination (MMSE).[22] The overall mean duration of disease was 4.25 years (SD = 1.71 years). Demographic features are summarized in table 1. All subjects completed an informed consent procedure approved by the Institutional Review Board at the University of Pennsylvania.

Materials. The subjects' task was to tell the story of the wordless children's picture book, *Frog, Where Are You?* [23] The story is given in its entirety in appendix E-1 (on the *Neurology* Web site at www.neurology.org). In brief, the story opens with a boy and his dog admiring a frog that they have in a large jar, as they prepare to go to bed for the night. When the boy and the dog are asleep, the frog climbs out of the jar and escapes through the window. Morning comes, and the boy and dog awaken to discover that the frog is gone. They set about looking for the frog, first in the boy's room, then in the nearby woods. They have a series of adventures involving encounters with a groundhog, a hive of bees, an owl, and a deer. They finally find their frog, only to discover that he is with a lady frog, and they have a brood of baby frogs. The boy and dog head for home with one of the babies, waving a cheerful goodbye to the frog family. We elected to study narrative discourse in this manner, rather than in free conversation, because the pictures would allow us to determine the accuracy of production relative to the intended target. We did not ask participants to tell us a fairy tale because the overlearned nature of such stories was expected to confound our ability to detect difficulties with narrative organization. We also did not ask subjects to describe a single scene such as the Cookie Theft picture because the material in a single scene is not rich enough to bring out a deficit in discourse performance. All print that appeared on the cover, back, and prefatory pages of the book was covered with heavy paper stock so that no print was visible.

The elements that are significant for assessing the narratives produced for this study are orientation, complicating action, and resolution. An analysis of the structure of the frog story yields seven episodes, each of which consists of an orientation, one or more elements of complicating action, and a resolution. These are the elements or events that a speaker must report to tell the story in full. The seven episodes comprise a total of 30 events. In some cases, two episodes overlap, when the boy is engaged in one activity and the dog is engaged in another. The events are defined in appendix E-1, which shows the overlap of episodes through the page number key. The coding of the narratives was performed using this structure of 30 events as the standard against which subjects' narrations were judged.

Narrative procedure. Each subject was asked to look through the book to become familiar with the story. When ready, the subject was asked to start at the beginning and tell the story as if he or she were reading it to a child. The story is told in a sequence of detailed drawings. All the subjects' narrations were digitally recorded. Thirty-three were recorded on a Macintosh Powerbook G3 laptop computer using the Macintosh external microphone (part no. 590-0670) and the computer program SoundEdit 16, version 2, with 16-bit recording at a sampling frequency of 44.1 K Hz. Six were recorded on a Dell Inspiron 2200 personal computer using the signal processing software Praat (Boersma and Weenink, version 4.2, 1992 to 2004) with 16-bit recording at a sampling rate of 22,050 Hz, using a Radio Shack omnidirectional lavaliere electret condenser microphone. Six were recorded on a Marantz PMD 670 digital recorder with 16-bit recording at a sampling frequency of 32 K Hz, using a Sennheiser MKE2 omnidirectional lavaliere condenser microphone.

The recordings of the narratives were transcribed in detail by trained transcribers using Praat. The transcription conventions used to capture the irregularities in patients' speech are defined in appendix E-2. All transcriptions were checked by two independent reviewers. The narratives were scored for discourse measures from the transcripts, referring to the original speech files as needed, by a linguist with expertise in narrative analysis.

The narratives elicited for the study were analyzed for features of coherence of the story as a whole and for maintenance of the theme. They were also analyzed for factual accuracy and for word-level difficulties that impinged on the overall communication of the story. The variables that were coded are as follows.

<u>Duration.</u> Duration was the total time spent on a speaker's narrative, from when he or she began telling the story until he or she indicated that the narrative was finished.

<u>Number of utterances.</u> An utterance was defined as a T-unit,[24] which consists of an independent clause and all clauses or phrases dependent on it. Therefore, a series of three independent clauses conjoined by *and* is counted as three utterances. A stretch of speech that formed an incomplete T-unit was also counted as an utterance.

<u>Number of words.</u> This count included all complete words, including repetitions.

<u>Lexical retrieval difficulty.</u> Five features that occur at the level of individual words were tabulated. They have a local effect in the discourse, and the accumulation of tokens of word-level difficulty contributes to the degree to which a speaker's story does or does not make sense overall. The features are 1) occurrence of word-finding difficulty, 2) a general noun in place of a specific one, 3) use of a wrong noun, 4) use of a wrong verb, and 5) a pronoun missing its antecedent.

<u>Content.</u> This was judged against the standard of the 30 events of content that were established independently of the speakers' narrations. An utterance that corresponded to a given event was scored as accurate if it conveyed the full content of the event with no contradictory content. It was scored as incomplete if it conveyed part of the essential content of the event. It was scored as an error if any aspect of what was said was factually inaccurate, and it was scored as content missing if there was no reference in the subject's narration to that event. Missing for content and missing for local connectedness are identical, as noted below.

<u>Action.</u> Each event in the story was coded for whether the interaction between actors and objects in the event was mentioned. Patients sometimes named the characters and objects that appeared in a scene but did not relate them to each other and tell what was happening. This measure overlaps those of content and local connectedness, but as a feature that affects coherence, it was deemed important to quantify it separately.

<u>Global connectedness.</u> The overall point of the story is that the boy and his dog search for the escaped frog, and they finally find him. Subjects were scored positively for global connectedness if they recognized that the frog found at end of the story is the same frog that figures in the opening pages, the one they have

been searching for. Subjects were scored as not showing global connectedness if they talked about the event in which the boy and dog come upon their frog but did not indicate that the frog had been present earlier in the story. Narrations were scored as missing for global connectedness if a subject did not mention that event in the narration.

Search theme. A second measure of overall coherence is maintenance of the theme of searching for the frog.[25] Search theme is scored from 0 to 4 by counting points accrued according to these criteria: one point for noting that the frog is missing, one point for noting that the boy is searching for the frog, one point for one or two further mentions of the search theme, and one point for any additional mentions of the search theme.

Local connectedness. An event was scored as locally connected if it presented some relation to the elements that preceded it. This is accomplished by rhetorical devices such as sequencing adverbials, pronominal reference to preceding events, reference by definite as opposed to indefinite determiners (given vs new information), and statements of cause and effect. An event was scored as not connected if no such devices were present and the reported event did not follow logically from the preceding utterances. It was scored as missing if there was no reference to that event; this is identical to missing content.

To test our hypothesis that elements of narrative production are related in part to executive resources such as planning, we administered a category naming fluency test, FAS. This task requires the subject to name as many words as he or she can think of that begin with the letter *F*, excluding proper nouns, numbers, and forms derived from a word already given. The task is repeated for the letters *A* and *S*. Performance on this measure is thought to reflect organization of a mental search through the lexicon, as well as lexical retrieval and speech fluency. We counted the number of unique and accurate responses provided for each target letter in 1 minute. Performance as measured by the total number of words produced on FAS is reported in table 1. We correlated performance on this task with elements of narrative production in patients with SOC/EXEC.

Imaging procedure. This procedure has been described in detail elsewhere.[10] Briefly, images were acquired by a GE Horizon Echospeed 1.5-T MRI scanner or a Siemens 3-T MRI scanner. Each study began with a rapid sagittal T1-weighted image to determine patient position. Next, high-resolution T1-weighted three-dimensional spoiled gradient echo images were acquired with TR = 35 milliseconds, TE = 6 milliseconds, slice thickness of 1.3 mm, flip angle of 30 °, matrix = 128 × 256, and in-plane resolution of 0.9 × 0.9 mm. The brain volumes were normalized by registration to the SPM99 T1 template of 305 averaged brains[26] using 12-parameters affine registration, nonlinear registration with 12 nonlinear iterations, and 7 × 8 × 7 basis functions. SPM99[27] was used to segment the brain volumes into four tissue types (gray matter, white matter, CSF, and other) with minimal inhomogeneity correction. The segmentation algorithm in SPM99 calculates a Bayesian probability for each voxel of each tissue group in the volume, based on a priori MRI information. A proportional analysis threshold was used to include only voxels with 40% or greater of gray matter. Implicit masking was used to ignore zeros, and global calculation was based on the mean voxel value. Using SPM99, the gray matter volume was smoothed with a 12-mm full width-half maximum gaussian filter to minimize individual gyral variations. SPM99 was used to perform a regression analysis relating cortical volume to the local connectedness measure of narrative organization in 9 of the patients with SOC/EXEC where imaging data were available. We set our statistical threshold at $p < 0.001$, and we accepted only clusters comprised of 100 or more adjacent voxels as significant, resulting in a p value that far exceeds the 0.05 level corrected for multiple comparisons.[28]

Results. The following extract illustrates effortful speech in the narrative of a PNFA patient (refer to appendix E-2 for a listing of transcription conventions):

1. The əhuh . . . d- boy . . . found . . um . . muskrat.
   The dog, looked up, at the beehive.
   The . . . muskrat . . . came outta fo-, the hole
   The . . /bΩəd/ {boy} looked in to də {the} tree hole . . . and (3.4 seconds) have foun' an owl.
   The boy fell down.

The overriding impression of the speech of patients with PNFA is that it is effortful and slow. Table 2 gives the speech rate in words per minute for the four subject groups, showing that the average rate of speech of patients with PNFA is less than one-third that of controls and approximately half that of the other two patient groups. A one-way ANOVA reveals an effect of group [$F(3,41) = 13.70$, $p < 0.001$]. The patients with PNFA are less fluent than each of the three other groups [controls: $t(18) = 9.37$, $p < 0.001$; SemD: $t(21) = 3.23$, $p < 0.005$; SOC/EXEC: $t(20) = 2.67$, $p < 0.01$]. In addition, the speech of both SemD and SOC/EXEC patients is slower than that of controls [SemD: $t(21) = 4.45$, $p < 0.001$; SOC/EXEC: $t(20) = 3.16$, $p < 0.01$]. The effortfulness of PNFA patients' speech is also reflected in the low mean length of utterance (MLU) in comparison with the other groups, also given in table 2. A one-way ANOVA shows a group effect [$F(3,41) = 4.40$, $p < 0.01$]. Patients with PNFA have a lower MLU than controls [$t(18) = 3.68$, $p < 0.01$] and patients with SOC/EXEC [$t(20) = 2.65$, $p < 0.05$].

Word-finding difficulty in the narratives of SemD patients is illustrated in the following two extracts:

2. And Sam went to sleep.
   And the dog went to sleep with him.
   And the frog was trying to get out of the . . what'd I call this . . . it's: a jar.

3. And he was sleeping with two animals
   And one animal woke him up
   Then he had his shoes or something on
   Another animal, the other animal got his head in a glass and a bottle, whatever it's called

Extract 2 illustrates naming difficulty in which the speaker with SemD searches for a particular word. In this case, the speaker is eventually able to come up with the desired form after a delay. In Extract 3, the SemD speaker does not attempt to find a precise word but rather uses the general term *animal* to refer to both the frog and the dog. These were the opening lines of this speaker's rendition of the story, and they illustrate an additional impairment in that the speaker uses the pronoun *he* with no antecedent, in place of producing *the boy*. Expressions such as *his shoes or something* and *a glass and a bottle, whatever it's called* express uncertainty or approximation in the use of the nouns that are so qualified. Finally, *glass* and *bottle* are inexact terms for the jar in the drawing, and *got his head in the bottle* is an imprecise formulation of what would properly be *got his head stuck in the bottle*.

Table 2 summarizes the word-level impairment seen in the four subject groups, particularly the patients with SemD, on five measures: word-finding difficulty as illustrated in Extract 2, and in Extract 3, a faulty verb, a wrong noun, production of a general noun, and a pronoun with no antecedent. Elderly control subjects have very little difficulty retrieving words to tell the story, but all three patient groups make errors or struggle to find appropriate words. A one-way ANOVA exhibits a group effect [$F(3,41) = 5.02$, $p < 0.01$]. All patient groups are impaired compared with controls [PNFA: $t(18) = 2.16$, $p < 0.05$; SemD: $t(21) =$

Table 2 *Mean (SD) performance on measures of discourse**

	Controls	PNFA	SemD	SOC/EXEC
Lexical retrieval and speech fluency				
Words per minute†	142 (24)	45 (22)‡	81 (28)‡§	90 (48)‡§
Mean length of utterance†	10.4 (1.8)	6.9 (2.4)‡	8.3 (2.3)	9.3 (2.4)
Impaired word-finding, frequency†	1.0 (1.2)	5.8 (6.9)	9.7 (6.7)‡	4.8 (4.5)
Event report accuracy, 30 events total				
Fully accurate†	25.1 (1.9)	8.5 (7.1)‡	9.8 (9.0)‡	13.4 (8.7)‡
Incomplete†	2.8 (1.4)	10.7 (1.9)‡	10.8 (5.7)‡	7.8 (3.2)‡
Errors†	0.7 (0.8)	1.0 (1.1)	2.9 (2.1)‡§	3.5 (3.3)§
Missing†	1.4 (1.5)	9.8 (7.1)‡	6.5 (4.5)‡	5.2 (5.4)
Frequency of "no action"	0.10 (0.32)	2.2 (3.3)	2.1 (3.9)	2.8 (4.4)
Story-level connectedness				
Global connectedness, 0–1†	1.0 (0.0)	0.70 (0.48)	0.62 (0.51)	0.25 (0.45)‡
Maintenance of search theme, 0–4	4.0 (0.0)	2.0 (1.7)‡	2.5 (1.8)	2.0 (1.6)‡
Local connectedness, 30 events total				
Connected	28.6 (1.5)	19.0 (7.5)‡	19.3 (9.1)‡	19.7 (9.9)
Not connected	0.0 (0.0)	1.2 (1.7)	4.2 (5.5)	5.1 (7.1)
Ratio of not-connected to present events	0.0 (0.0)	0.07 (0.12)	0.21 (0.30)	0.24 (0.34)

* All values significant at least at the $p < 0.01$ level (see text for details).
† Univariate F ratio comparing groups.
‡ Group differs from controls.
§ Group differs from progressive nonfluent aphasia (PNFA) group.

SemD = semantic dementia; SOC/EXEC = disorder of social comportment and executive functioning.

4.06, $p < 0.001$; SOC/EXEC: t(20) = 2.52, $p < 0.05$]. Patients with SemD also are impaired compared with patients with SOC/EXEC [t(23) = 2.20, $p < 0.05$].

An example of content misinterpretation in the narrative of a SOC/EXEC patient is as follows:

4. Uh, and the little boy looks out of his bedroom window and calls for the frog.
   And the dog's got his head in a jar, in the frog's jar, jar.
   The little boy is still at his bedroom window, looking at the dog, trying to get his head out of the frog's jar
   Well the jar is broken
   And the dog does get his head out of the jar that way
   And the little boy is glad of that.
   He's huggin' the, dog.

In terms of lexicon and sentence structure, the speech of SOC/EXEC patients is much less deviant than that of patients with PNFA and SemD. However, it would not be taken to be normal. Extract 4 is grammatically well formed and reports the actions of the participants accurately, until the end of the extract. The corresponding page of the book shows the boy holding the dog, and the dog is clearly happy, licking the boy's face, glad to be released from the jar that had been stuck on his head. The boy, however, is frowning, with eyebrows knitted, clearly displeased that the dog has broken the jar. This SOC/EXEC patient speaks well, but she errs in reporting the content by misinterpreting the scene.

Table 2 shows that patients with FTD report fewer accurate events and more incomplete events than controls and that patients with SOC/EXEC are as impaired as the progressive aphasics on these measures. Control subjects average full accuracy on the majority of events in the story. A one-way ANOVA reveals an effect of group [F(3,41) = 10.53, $p < 0.001$], which is attributable to the poor performance of patients with FTD relative to controls [PNFA: t(18) = 7.16, $p < 0.001$; SemD: t(21) = 5.23, $p < 0.001$; SOC/EXEC: t(20) = 4.17, $p < 0.001$]. The controls' rate of incomplete events is approximately 10%. The production of incomplete events follows the same pattern as that of fully accurate reports: an effect of group is found [F(3,41) = 10.82, $p < 0.001$], all patient groups are impaired relative to controls [PNFA: t(18) = 10.63, $p < 0.001$; SemD: t(21) = 4.29, $p < 0.001$; SOC/EXEC: t(20) = 4.55, $p < 0.001$], and SOC/EXEC patients without aphasia are as impaired as PNFA patients and SemD patients.

Table 2 also shows that patients with FTD make more errors when reporting story events than do controls and that patients with SOC/EXEC are as impaired as those with progressive aphasia. A one-way ANOVA shows an effect of group [errors: F(3,41) = 4.64, $p < 0.01$]. Relative to controls, more factual errors are made by patients with SemD [t(21) = 3.15, $p < 0.01$] and patients with SOC/EXE [t(20) = 2.62, $p < 0.05$]. Patients with SemD [t(21) = 2.64, $p < 0.05$] and patients with SOC/EXEC [t(20) = 2.46, $p < 0.05$] also produce more factual errors than patients with PNFA. A high frequency of missing elements is also found in the discourse of all FTD patient groups compared with controls [F(3,41) = 4.79, $p < 0.01$]. Relative to controls, nonaphasic patients with SOC/EXEC [t(20 = 2.17, $p < 0.05$] are as impaired as patients with PNFA [t(18) = 3.65,

$p < 0.005$] and patients with SemD [t(21) = 3.47, $p < 0.005$].

The following extract illustrates a particular kind of content error that is common in the narratives of SOC/EXEC patients; this is the description of the content of a picture without mention of the actions in the picture that further the story line:

5. (a) The boy is asleep in his bed.
The frog is in a jar.
And his {chuckle} . . his . . his boots are on the floor, nex-next to his uh . . . next to his shirt.
(b) Boy's in bed (2 seconds) next to his dog.
His boots are on the floor . .
And so are his sandals . . and an empty jar, and his shirt.

The first three lines, 5(a), refer to the second page of the story, in which the boy is asleep in bed and the dog is curled up on the bedspread at the foot of the bed. The frog is in the process of climbing out of the jar; his body and one leg are out, while the other leg is dangling down inside the jar. The next three lines, 5(b), refer to the following page. There, it is clearly daytime, with sun streaming in through the window, and the boy is crouched on top of the bedcovers, staring at the empty jar, with a look of dismay on his face. The dog, too, is standing up on the bed, looking woefully at the jar. The speaker first of all does not make any connection between the picture described in 5(a) and the preceding scene, the beginning of the story, in which the boy and dog are ready for bed but are on the floor, admiring the frog. A connection could be made to this scene by saying, "The boy *has gone* to sleep" or "The boy *goes* to sleep." In saying instead, in the first line of Extract 5, that "The boy *is* asleep," the narrative describes a simple state of being rather than a flow of events. In addition, 5(a) does not describe any action at all, although the action of the frog climbing out of the jar is absolutely clear, and it is crucial to the story. 5(b) likewise fails to convey the event of the next page, the boy's discovery that the frog has disappeared and his stunned surprise at this development; the speaker simply names several objects that appear in the scene. This failure to describe the action of the story line, in favor of simply describing (or only naming) the characters and objects in the picture, is a common feature of SOC/EXEC patients' narratives. The frequency of such no-action descriptions is presented in table 2. Control subjects exhibit this ineffectiveness of narration rarely, whereas all three groups of FTD patients show it with some frequency. Progressive aphasics may have word-finding difficulty, but only patients with SOC/EXEC exhibit the no-action feature more frequently than controls [t(20) = 1.97, $p = 0.06$].

Global connectedness is assessed by determining whether the speaker, upon reaching the end of the story, recognizes that the frog found by the boy and his dog is the same frog that figures in the opening pages, the one they have been searching for. The scene of actual discovery is preceded by a sequence of pictures in which, first, the boy cocks his ear, having heard a sound that attracts his attention. Then, the boy and dog lean over a big log, while the reader sees only their backs. The climax comes in the next picture, in which the reader, along with the boy and his dog, see the original frog, with a girl frog affectionately leaning against him. In the following picture, a cluster of baby frogs emerges from the brush. One control subject described the scene thus:

6. The sound must be coming from behind the log.
They both lean over the log.
What do they see?
They see Flip the frog and his girlfriend Florrie.
Florrie and Flip, there they are.
There they are with their little froglets!

This description, told with strongly affective prosody, may be compared with that of a SOC/EXEC patient who fails to recognize any connection with an earlier part of the narrative:

7. Dog—or boy's . . over log
Dog's over the log too
Um . . . they're on the log
See two frogs
See the mom and . . . dad and a mom frog
And you got one, two, three, four, five . . seven little—eight little toads.

Table 2 shows poor global connectedness in patients with SOC/EXEC, i.e., their difficulty recognizing at the end of the story that the boy and dog have found the frog that was with them at the beginning. There is an overall effect of group [$\chi^2(3) = 13.40$, $p < 0.005$]. None of the controls misses this, the overall point of the story. The aphasic groups succeed in making the connection approximately two-thirds of the time. However, patients with SOC/EXEC make the connection between the frog at the beginning and the frog at the end in only one quarter of the cases. The success of patients with SOC/EXEC is less than that of controls [$\chi^2 = 11.86$, $df = 1$, $p < 0.001$], patients with PNFA [$\chi^2(1) = 4.46$, $p < 0.05$], and patients with SemD [$\chi^2(1) = 3.38$, $p = 0.06$]. Patients with SemD also perform worse than controls on this measure [$\chi^2(1) = 4.92$, $p < 0.05$]. Moreover, the difficulty of patients with SOC/EXEC on global connectedness correlates with their poor scores on the FAS test [$r(10) = 0.69$; $p < 0.05$], and this correlation was seen only in patients with SOC/EXEC.

Another measure of coherence that is found to be impaired in patients with SOC/EXEC is maintenance of the search theme throughout the story. As summarized in table 2, there is an effect of group [F(3,41) = 4.07, $p < 0.05$]. Relative to controls, nonaphasic patients with SOC/EXEC [t(20) = 3.95, $p < 0.005$] were as impaired as patients with PNFA [t(18) = 3.72, $p < 0.005$] and patients with SemD [t(21) = 2.68, $p < 0.05$].

Table 2 also shows that patients with SOC/EXEC are the most impaired group in terms of their local connectedness. Controls occasionally omit events (see content errors for missing events), but they do not make the outright error of failing to connect an event that they mention to the one preceding. Patients with FTD differ from controls in failing to connect approximately one-third of their mentioned events [F(3,41) = 3.54, $p < 0.05$]. Relative to controls, the patients with SOC/EXEC [t(20) = 2.82, $p < 0.05$] are as impaired as the patients with progressive aphasia [PNFA: t(18) = 3.99, $p < 0.01$; SemD: t(21) = 3.18, $p < 0.01$]. In terms of outright errors of connectedness, patients with SemD are more impaired than patients with

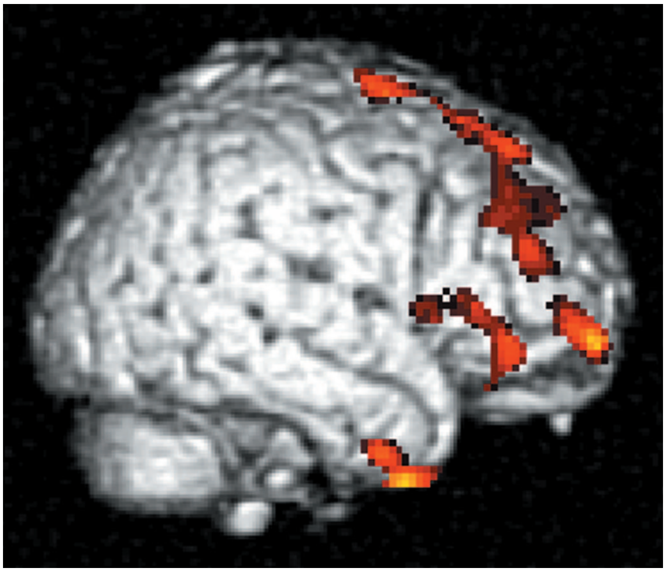

Figure. Lateral view illustrating areas of cortical atrophy significantly related to the local connectedness measure of narrative organization.

PNFA, whereas patients with SOC/EXEC exhibit the poorest performance. This is revealed in the ratio of unconnected events to the total number of events that are present in the speakers' narratives, given in table 2. Less than 10% of the PNFA patients' mentioned events lack local connectedness (which does not differ significantly from controls), whereas approximately one-fifth of SemD patients' mentioned events [$t(21) = 2.22$, $p < 0.05$] and one quarter of SOC/EXEC patients' mentioned events [$t(20) = 2.23$, $p < 0.05$] are not locally connected, differing from controls. Only patients with SOC/EXEC exhibit an inverse relation of not-connected events with FAS performance [$r(10) = -0.69$; $p < 0.05$].

We used a regression analysis to relate local connectedness to cortical atrophy in nine SOC/EXEC patients with available MRI. The figure illustrates the significant correlations. As noted in table 3, we find that local connectedness is related to several right hemisphere regions, including dorsolateral prefrontal, inferior frontal, superior frontal, and anterior temporal cortical areas.

Discussion. We asked patients with FTD to tell a story from a sequence of pictures. Despite the superficial simplicity of this task, there are many steps involved in producing a narrative. Traditional assessments of language production measure the ability to perform such tasks as recognizing and naming the objects and actions of a story and linking these words in syntactically well-formed sentences. In the current assessment of narrative, we focused on the patients' additional need to draw on discourse resources to express the larger ideas that form a story. The patients had to infer the point of each picture and its contribution to the story as a whole. They also had to recognize the setting of the stage for each event, the critical participants and actions, and the outcome, while keeping in mind the overall point of the story. They had to connect the events locally by organizing the interpretation of each event to relate it to the ones preceding it, and, finally, they had to maintain the theme of searching for the lost frog and the fact that a resolution was reached. The task is complex, and this is the basis for communicating meaningful messages in everyday conversation.

Despite the importance of discourse for social interaction and communication, investigations of narrative discourse in neurodegenerative diseases have been rare. In one report, patients with Alzheimer disease (AD) were found to be impaired on what was termed "macro-level processing" but not on the "microlinguistic" levels of syntax and lexicon, whereas stroke patients showed the converse pattern of impairment.[5] Another study found that patients with AD are impaired at both the levels of "gist" (overall interpretation and inferencing) and "detail" (accuracy of content).[29] Impairments of semantic processing during expository discourse have been observed in both AD and vascular dementia.[6] In a longitudinal study of a small group of progressive aphasic patients examining expository and conversational speech samples, three out of four patients exhibited a decline in their grammar, with increasingly impaired retrieval of verbs and closed-class elements over time. The fourth patient showed declining word-

Table 3 *Regression analysis evaluating the relationship between cortical atrophy and the local connectedness measure of narrative organization in patients with SOC/EXEC*

	Coordinates*				
Anatomic region (Brodmann area)	x	y	z	No. of voxels	Z score
Right inferior frontal (47, 45)	38	37	−7	481	3.58
Right dorsolateral prefrontal (10)	44	44	18	321	3.14
Right polar prefrontal (10)	32	54	−3	270	3.18
Right superior frontal (6)	24	5	64	380	3.19
Right anterior temporal (38, 21)	32	8	−37	336	3.80
Left superior frontal (6)	−10	5	66	416	3.75

* Peak anatomic coordinates refer to the neuroanatomic atlas of Talairach and Tournoux.[46]

SOC/EXEC = disorder of social comportment and executive functioning.

retrieval ability, with relatively little grammatic difficulty.[7] Descriptions of the Cookie Theft scene in SemD showed an impairment in the use of low frequency nouns, and three patients with SemD who were studied longitudinally were particularly impoverished in their use of low-frequency nouns that are imageable.[30]

In the current study, control subjects are quite good at carrying out the complex set of tasks needed to produce a coherent narrative. All three groups of FTD patients, by comparison, show impairments in performing this task. Moreover, the nature of the deficit in each subgroup of FTD patients is relatively distinct. Patients with SemD seem to be impaired at retrieving the words needed to express a narrative. Patients with PNFA are quite effortful in their speech, which results in sparse narratives. The performance of patients with SOC/EXEC is striking in that these nonaphasics are the most impaired patients at using the principles of discourse to produce a narrative. Although they find the individual words to describe a picture and the syntax to combine the words into sentences, their narratives lack the elements of local and global connectedness needed to unite the elements of the story into a coherent whole. Local and global connectedness scores are correlated with performance on an independent measure of mental search. A crucial role thus may be played by executive resources such as planning and organization that are compromised in patients with SOC/EXEC.[13-15] It is this ability to organize that is impoverished in the day-to-day speech of patients with SOC/EXEC, presumably contributing to the frustration frequently voiced by their families over everyday communication. Moreover, the organizational deficit in SOC/EXEC patients' narratives is related to their right frontotemporal disease. This parallels the findings of studies of nonaphasic stroke patients with a discourse impairment, which also implicate the right hemisphere.[11,12] In sum, we hypothesize that distinct patterns of narrative impairment reflect essential clinical features of each FTD phenotype. We review each subgroup of patients with FTD in turn below.

For patients with SOC/EXEC, impairment emerges at a linguistic level higher than that of producing individual words or constructing sentences. They are compromised in their ability to relate successive events to one another, and they have difficulty relating the end of the story to the beginning. This is forcefully demonstrated by the poor performance of patients with SOC/EXEC on the measure of global connectedness, the expression of the resolution that justifies the telling of the story. On this measure, patients with SOC/EXEC are significantly impaired compared with controls and also compared with PNFA and SemD patients. They are also significantly impaired in their ability to maintain the search theme and local connectedness throughout the narrative. Such difficulty in maintaining the organization of the narrative is abundantly evident among the patients with SOC/EXEC, even though they are not aphasic.

We hypothesized that these measures depend in part on executive resources such as organization, planning, and working memory, which are needed to maintain the thread of a narrative, to relate one event to another, to infer relations of cause and effect, and to grasp the overall point or goal of a story.[1] These resources need not necessarily be specific to language processing. An individual draws on these resources for carrying out a multitude of routine actions which involve deciding on a goal, determining the steps needed to accomplish the goal, taking into account the necessary order of the steps, and then following through with carrying out the steps, keeping track of the steps themselves and the progress from step to step until the conclusion is reached. Just as they are required for getting dressed and preparing breakfast, they also seem to be required for recounting a story.[31,32] Indeed, we have observed significant difficulty among patients with SOC/EXEC in relating the organization of events in a script, and this is found to be more compromised than their ability to report content accurately.[33] Therefore, the difficulty of patients with SOC/EXEC in producing coherent, organized discourse parallels their difficulties in carrying out organized routine tasks of daily living.[8-10] The correlations of performance on the FAS test with measures of global connectedness and local connectedness in patients with SOC/EXEC are consistent with the hypothesis that these patients have difficulty bringing executive resources to bear during narrative production, and this interferes with their ability to tell a coherent story.

Additional evidence that the narrative deficit in patients with SOC/EXEC is not related to aphasic deficits in lexical retrieval and syntax comes from the imaging study relating a measure of narrative organization to cortical atrophy in these patients. We find that discourse difficulty in patients with SOC/EXEC is related to disease in the right hemisphere, particularly in the right frontal and temporal cortices. This is consistent with previous work on language difficulties of nonaphasic patients after a right hemisphere stroke, where a deficit in discourse also has been noted.[11,12] fMRI studies of narrative in healthy adults show activation of frontal and temporal regions.[34] In particular, activation of right prefrontal and temporal regions is associated with the extraction of the overall meaning of long sentences and short stories.[35,36] Lesion studies emphasize the role of right frontal insult in the appreciation of large scale structures such as scripts,[37-39] a deficit also seen in patients with SOC/EXEC.[33] Previous studies of patients with SOC/EXEC have shown cortical atrophy in these right frontal and temporal regions, and the observations of the current study confirm the functional significance of these brain regions for discourse.[8-10] Independent evidence that these frontotemporal brain regions contribute to executive resources comes from fMRI studies of healthy adults that show activation of these areas when performing planning and working memory tasks.[40-42]

We surmise that the narrative deficit in patients with SOC/EXEC is not due to an episodic memory limitation because these patients do not link even successive events of a story that do not depend on memory. Similarly, their deficit is not easily attributed to a semantic memory impairment because they have no difficulty describing a picture. Instead, patients with SOC/EXEC have difficulty telling a story that makes sense because they cannot understand or express the logical connection of one event to another. This difficulty is exemplified in the relatively high frequency of no-action descriptions of scenes in the book. The patients with SOC/EXEC frequently enumerate the components of a scene in the story but do not describe the intended action. If a given scene does not seem to be connected to the one preceding, there is little to say about it other than to describe the characters and objects that are present. A factor that potentially contributes to this could be the disorder of social behavior seen in patients with SOC/EXEC. The consequence of this disorder is expressed in a sequence of utterances such as Extract 4. To the extent that SOC/EXEC patients' misinterpretations are limited to the mood indicated by a facial expression, these patients can be said to have a category-specific semantic impairment for social knowledge[39,43] that interferes with the accurate production of a narrative. However, a social deficit cannot explain their pervasive difficulty with local connectedness throughout the story regardless of event content or their difficulty with the overall point of the story (global connectedness).

Patients with PNFA have significantly reduced fluency and produce abbreviated utterances in their speech.[7] They attain low scores on all measures relating to the volume of production, including words per minute, total number of words, and MLU. A reduced MLU is often associated with impoverished sentence-level grammatic production, as noted previously in PNFA patients' descriptions of a single complex picture.[7] We too observed some evidence of difficulty with sentence-level grammar in the omission and incorrect use of grammatic forms, as in Extract 1. The effortful nature of these patients' storytelling is also due in part to apparent difficulty articulating what they plan to say, which is reflected in their frequent speech errors. Regardless of the basis for the impoverished speech production in PNFA, one consequence is a high rate of missing elements. While patients with PNFA perform poorly on local connectedness because they have a high rate of missing elements, they exhibit local connectedness at the same rate as controls for those elements that are present. This contrasts with the performance of patients with SOC/EXEC who have missing elements, because the missing elements of patients with SOC/EXEC contribute to their poor connectedness scores. The nonfluent speech of patients with PNFA may likewise explain their limited success at maintaining the search theme. However, they are not significantly different from controls on global connectedness, the measure of their ability to understand and express the overall point of the story. They comprehend the story well, and so their performance on higher-level discourse measures of connectedness is quite good when they are able to describe an event. Therefore, patients with PNFA have less difficulty than SOC/EXEC patients with the cohesiveness of the narrative, but they have significantly greater difficulty with fluent speech production.

In SemD, the most distinctive disability is that of impoverished lexical access and meaning. Patients with SemD make significant mistakes in lexical retrieval, failing to mention items for which they cannot find a word. They also produce semantic substitutions or the wrong words for objects and provide imprecise superordinate terms such as *animal* or *thing* in place of a more informative object name that would be appropriate in a narrative. These characteristics of SemD speech, also noted in more structured measures such as confrontation naming and the description of a single picture, such as the Cookie Theft scene, may be related to a fundamental impairment of semantic knowledge.[30,44] Lexical access and retrieval difficulty in SemD contributes to a paucity of content, resulting in a high rate of incomplete and missing content elements. For this patient group, reduced content is associated with borderline impairment of local and global connectedness, along with modest difficulty on the search theme measure.

As has been found for stroke aphasics,[5,45] the two groups of progressive aphasic patients with FTD studied here do not perform poorly at the level of discourse per se. Rather, their nondiscourse impairments account for their difficulties in telling the story effectively, because problems such as effortfulness of speech and poor lexical access ultimately interfere with their production of the elements of the story. These two features—effortful speech and poor lexical access—distinguish the two aphasic groups from each other. Impairments that truly affect the higher level of discourse are less prominent than in the nonaphasic FTD patients with a SOC/EXEC clinical profile.

References

1. Mar RA. The neuropsychology of narrative: story comprehension, story production, and their interrelation. Neuropsychologia 2004;42:1414–1434.
2. Labov W, Waletzky J. Narrative analysis: oral versions of personal experience. In: Helm J, ed. Essays on the verbal and visual arts: Proceedings of the 1966 Annual Spring Meeting of the American Ethnological Society. Seattle: University of Washington Press, 1967:12–44.
3. Grossman M. Frontotemporal dementia: a review. J Int Neuropsychol Soc 2002;8:564–583.
4. Snowden JS, Neary D, Mann DM. Fronto-temporal lobar degeneration: fronto-temporal dementia, progressive aphasia, semantic dementia. 1st ed. New York: Churchill Livingstone, 1996.
5. Glosser G, Deser T. Patterns of discourse production among neurological patients with fluent language disorders. Brain Lang 1990;40:67–88.
6. Vuorinen E, Laine M, Rinne J. Common pattern of language impairment in vascular dementia and in Alzheimer disease. Alzheimer Dis Assoc Disord 2000;14:81–86.

7. Thompson CK, Ballard KJ, Tait ME, Weintraub S, Mesulam M. Patterns of language decline in non-fluent primary progressive aphasia. Aphasiology 1997;11:297–331.
8. Grossman M, McMillan C, Moore P, et al. What's in a name: voxel-based morphometric analyses of MRI and naming difficulty in Alzheimer's disease, frontotemporal dementia, and corticobasal degeneration. Brain 2004;127:628–649.
9. Rosen HJ, Gorno-Tempini ML, Goldman WP, et al. Patterns of brain atrophy in frontotemporal dementia and semantic dementia. Neurology 2002;58:198–208.
10. Williams GB, Nestor PJ, Hodges JR. Neural correlates of semantic and behavioural deficits in frontotemporal dementia. Neuroimage 2005;24: 1042–1051.
11. Gardner H, Brownell HH, Wapner W, Michelow D. Missing the point: the role of the right hemisphere in the processing of complex linguistic materials. In: Perecman E, ed. Cognitive processing in the right hemisphere. New York: Academic Press, 1983:169–191.
12. Joanette Y, Goulet P. Narrative discourse in right-brain-damaged right-handers. In: Joanette Y, Brownell HH, eds. Discourse ability and brain damage: Theoretical and empirical perspectives. New York: Springer Verlag, 1990.
13. Boone K, Miller BL, Lee A, Berman N, Sherman D, Stuss D. Neuropsychological patterns in right versus left frontotemporal dementia. J Int Neuropsychol Soc 1999;5:616–622.
14. Kramer JH, Jurik J, Sha SJ. Distinctive neuropsychological patterns of frontotemporal dementia, semantic dementia, and Alzheimer's disease. Cogn Behav Neurol 2003;16:211–218.
15. Rahman S, Sahakian BJ, Hodges JR, Rogers RD, Robbins TW. Specific cognitive deficits in mild frontal variant frontotemporal dementia. Brain 1999;122:1469–1493.
16. The Lund and Manchester Groups. Clinical and neuropathological criteria for frontotemporal dementia. J Neurol Neurosurg Psychiatry 1994;57:416–418.
17. McKhann G, Trojanowski JQ, Grossman M, Miller BL, Dickson D, Albert M. Clinical and pathological diagnosis of frontotemporal dementia: report of a work group on frontotemporal dementia and Pick's disease. Arch Neurol 2001;58:1803–1809.
18. Davis KL, Price C, Moore P, Campea S, Grossman M. Evaluating the clinical diagnosis of frontotemporal degeneration: a re-examination of Neary et al, 1998. Neurology 2001;56:A144–A145.
19. Grossman M, Ash S. Primary progressive aphasia: a review. Neurocase 2004;10:3–18.
20. Price C, Davis KL, Moore P, Campea S, Grossman M. Clinical diagnosis of frontotemporal dementia (FTD). Neurology 2001;56:A176.
21. Neary D, Snowden JS, Gustafson L, et al. Frontotemporal lobar degeneration: a consensus on clinical diagnostic criteria. Neurology 1998;51: 1546–1554.
22. Folstein MF, Folstein SF, McHugh PR. "Mini Mental State": A practical method for grading the cognitive state of patients for the clinician. J Psychiatr Res 1975;12:189–198.
23. Mayer M. Frog, where are you? New York: Penguin Books, 1969.
24. Hunt KW. Grammatical structures written at three grade levels. Champaign, IL: National Council of Teachers of English, 1965.
25. Reilly J, Losh M, Bellugi U, Wulfeck B. "Frog, where are you?" Narratives in children with specific language impairment, early focal brain injury, and Williams syndrome. Brain Lang 2004;88:229–247.
26. Evans AC, Collins DL, Mills SR, Brown ED, Kelly RL, Peters TM. 3D statistical neuroanatomical models from 305 MRI volumes. In: Klaisner LA, ed. Nuclear Science Symposium and Medical Imaging Conference, Volumes 1-3, IEEE Conference Record. San Francisco, CA, 31 October-6 November 1993. Piscataway, NJ: IEEE Service-Center, 1993:1813–1817.
27. Frackowiak RSJ, Friston KJ, Frith CD, Dolan RJ, Mazziotta JC. Human brain function. San Diego: Academic Press, 1997.
28. Forman SD, Cohen J, Fitzgerald M, Eddy WF, Mintun M, Noll DC. Improved assessment of significant activation in functional magnetic resonance imaging (fMRI): use of a cluster-size threshold. Magn Res Med 1995;33:636–647.
29. Chapman SB, Zientz J, Weiner M, Rosenberg R, Frawley W, Burns MH. Discourse changes in early Alzheimer disease, mild cognitive impairment, and normal aging. Alzheimer Dis Assoc Disord 2002;16:177–186.
30. Bird H, Lambon Ralph MA, Patterson K, Hodges JR. The rise and fall of verb frequency and imageability: noun and verb production in semantic dementia. Brain Lang 2000;73:17–49.
31. Stine EAL, Wingfield A. How much do working memory deficits contribute to age differences in discourse memory? Eur J Cogn Psychol 1990; 2:289–304.
32. Titone D, Prentice KJ, Wingfield A. Resource allocation during spoken discourse processing: effects of age and passage difficulty as revealed by self-paced listening. Mem Cognit 2000;28:1029–1040.
33. Cosentino S, Chute D, Libon DJ, Grossman M. How does the brain represent scripts? A study of executive processes and semantic knowledge in dementia. Neuropsychology (in press).
34. Kemeny S, Ye FQ, Birn RM, Braun AR. Comparison of continuous overt speech fMRI using BOLD and arterial spin labeling. Hum Brain Mapp 2005;24:173–183.
35. Kircher TTJ, Brammer M, Tous Andreu N, Williams SCR, McGuire PK. Engagement of right temporal cortex during processing of linguistic context. Neuropsychologia 2001;39:798–809.
36. Xu J, Kemeny S, Park G, Frattali C, Braun A. Language in context: emergent features of word, sentence, and narrative comprehension. Neuroimage 2005;25:1002–1015.
37. Crozier S, Sirigu A, Lehericy S, et al. Distinct prefrontal activations in processing sequence at the sentence and script level: an fMRI study. Neuropsychologia 1999;37:1469–1476.
38. Sirigu A, Zalla T, Pillon B, Grafman J, Dubois B, Agid Y. Planning and script analysis following prefrontal lobe lesions. In: Grafman J, Holyoak KJ, Boller F, eds. Structure and functions of the human prefrontal cortex. New York: New York Academy of Sciences, 1995:277–287.
39. Wood JN, Romero SG, Makale M, Grafman J. Category-specific representations of social and nonsocial knowledge in human prefrontal cortex. J Cogn Neurosci 2003;15:236–248.
40. Botvinick MM, Braver TS, Barch DM, Carter CS, Cohen JD. Conflict monitoring and cognitive control. Psychol Rev 2001;108:624–652.
41. Ramnani N, Owen AM. Anterior prefrontal cortex: Insights into function from anatomy and neuroimaging. Nat Rev Neurosci 2004;5:184–194.
42. Smith EE, Marshuetz C, Geva A. Working memory: findings from neuroimaging and patient studies. In: Grafman J, ed. Handbook of neuropsychology. Vol. 7. New York: Elsevier Science, 2002:55–72.
43. Wood JN, Grafman J. Human prefrontal cortex: Processing and representational perspectives. Nat Rev Neurosci 2003;4:139–147.
44. Lambon Ralph MA, McClelland JL, Patterson K, Galton CJ, Hodges JR. No right to speak? The relationship between object naming and semantic impairment: neuropsychological evidence and a computational model. J Cogn Neurosci 2001;13:341–356.
45. Ulatowska HK, Freedman-Stern R, Doyel AW, Macaluso-Haynes S. Production of narrative discourse in aphasia. Brain Lang 1983;19:317–334.
46. Talairach J, Tournaux P. Co-planar stereotaxic atlas of the human brain. New York: Thieme, 1988.