In 1907 Alzheimer described a demented woman who “frequently used perplexing phrases...some paraphrastic expressions” (milk-pourer instead of cup) and suffered from a significant language comprehension deficit. Language disorders continue to be one of the most obvious and well-studied symptoms of dementia and are particularly pronounced in dementia of the Alzheimer type (DAT). The incidence of language impairment in dementia is estimated to be between 88 and 95 percent, and is close to 100 percent in DAT. This chapter will (1) describe the language disorder associated with various stages of DAT; (2) review several prominent research questions about the origin and interpretation of these language disorders; (3) discuss clinical implications of this research for diagnosis and treatment of language disorders in DAT; and (4) sketch out important areas of future research.

Overview of Language in DAT

The many descriptions of language in dementia that have appeared over the past decade confirm and expand Alzheimer’s original observation of word-finding deficits, paraphasias, and comprehension impairment. It should be noted that Alzheimer patients have little or no peripheral weakness or incoordination, and have no obvious motor speech deficits: control of phonation, articulation, and resonance remains intact until the latest stages. In addition, the ability to arrange words into grammatical sentences appears to be relatively spared throughout the course of the disease. The communication problems of this population are largely confined to the two aspects of language: lexicon (word knowledge) and pragmatics, the use of contextually appropriate language. As might be expected with a degenerative process, the deficits change over time, and each phase of the disease is characterized by a distinct profile of language deficits.*

EARLY-STAGE DEFICITS

The earliest language deficit observed in DAT is anoma. DAT patients have difficulty coming up with words in structured tasks such as word-list generation (e.g., name as many animals as you can) as well as in elicited narratives and spontaneous conversation. Semantically empty words (e.g., thing, stuff, do) are scattered throughout the DAT patient’s utterances in place of content words, thereby maintaining fluency and sacrificing informational content. Language comprehension for simple, structured, concrete material appears intact during these early stages. However, comprehension of abstract language that does not rely on meaning of single words in syntactic structures, but rather requires inference (e.g., Rome wasn’t built in a day; She took a sudden turn for the worse), is poor even in the earliest stages of DAT. Also, early on, patients have a difficult time generating spontaneous language via writing, although the mechanics of writing and

*Throughout this chapter, the terms “early/mild,” “moderate,” and “severe/late” will be used to describe the non-discrete stages associated with the 5 to 15 year course of DAT, which begins with mild cognitive symptoms and ends in a persistent vegetative state.
reading remain intact. At this point, DAT patients can communicate sufficiently for most social situations, although they may not be able to follow complex conversations and may tend to digress or repeat themselves. Although the DAT patient is often initially aware of his or her own language deficits, this awareness appears to wane by the end of the early phase.

A sample of a narrative description (from Kempler) of the cookie theft picture from The Boston Diagnostic Aphasia Examination (Fig. 7.1) illustrates some of these points. Note particularly the repetition of ideas (Momma’s washing dishes), empty words (somebody’s doing), paraphasias (single pencil), difficulty with appropriate use of pronouns (My teacher... they...), and a hint of awareness of the difficulty (My teacher should be here... ). Despite these difficulties, this patient is able to describe the major events and participants in the picture.

Examiner
I want you just to tell me what’s going on in this picture.

Alzheimer patient
Momma’s washing dishes and children are taking jam or jelly or something from the jar, and the little fella’s about ready to fall on the floor. And Momma’s washing dishes.

Examiner
Good. Anything else?

Alzheimer patient
Well, the overflow of the water.

MODERATE-STAGE DEFICITS

By the moderate stages of DAT, patients begin to have more difficulty with both production and comprehension of language. In production, anomia worsens and word-finding deficits are made more obvious by copious substitutions of empty words and circumlocutions for information-bearing nouns and verbs. The utterances of moderate DAT patients are often difficult to follow because of pragmatic deficits, including poor topic maintenance and poor use of pronouns. It is somewhat surprising that in the context of these pronounced discourse deficits, certain other discourse skills, such as turn-taking in conversation, remain undisturbed. Comprehension for complex material (e.g., sequential instructions) is often impaired by this stage. Although the mechanics of reading aloud and writing remain unimpaired, deficits in producing well-formed coherent writing and reading for comprehension parallel problems observed in auditory-spoken language. In the moderate stages, DAT patients'
conversations become difficult to follow, and the patients may withdraw slightly from social situations in which communication demands may occur. They often appear unaware of their communicative deficits at this point.

The more severe language deficits associated with this stage are illustrated by another narrative description of the cookie theft picture. At this point, the empty words and paraphasias render the narrative largely uninterpretable. The patient's use of descriptive phrases that have no direct relationship to the picture (canned goods, politically well known) elicit several requests for clarification from the examiner. However, the patient appears unable to clarify the narrative. Notice the preservation of sentence structure, including some syntactic complexities (e.g., "apparently because he has a tendency to be falling").

Examiner
I just want you to describe what's going on in this picture. Tell me what's going on here.

Alzheimer patient
This picture incorporates a certain amount of coming and going, and cheating, and doing the things in the world...[pause]...planning some for uh money on the floor, not worrying about it. Falling onto the ground.

Examiner
Speak a little louder.

Alzheimer patient
Oh that he has the ability to—she has the ability to change the timing and then and then the wind is blowing in the wind, blowing in the floor. Wind is blowing on the canned goods.

Examiner
The wind is blowing on the canned goods?

Alzheimer patient
Yeah, splattering.

Examiner
What she doing (point to woman at sink)?

Alzheimer patient
She is not only losing, doing, washing her payroll there, but also being careful of the child, apparently because he has a tendency to be falling this, slipping. And then there's a child slipping again, or more normally.

Examiner
More normally?

Alzheimer patient
Yeah, politically well known.

Examiner
Politically well known. What's going on over here (point to stool)?

Alzheimer patient
Well, it seems to be that they're trying to, filling up and ideal of it inside there...[pause]...things that we eat...play with and maybe get hurt. Candy or something.

LATE-STAGE DEFICITS

By the later stages of the disease, verbal production becomes uninterpretable because of paraphasias (word and sound substitutions) and lack of coherence. Late in the course of the disease, dysarthria may impair speech intelligibility. Eventually, the patients manifest echolalia (repetition of others), palilalia (repetition of self), and muteness. At this point, comprehension is impaired in all modalities, even for single words, and the patients are no longer successfully participating in social interaction through language or any other communicative modalities.

INTERPRETING LANGUAGE DEFICITS IN DAT

A few important observations about language in DAT must be mentioned before proceeding. First, there are language impairments that do not occur in DAT. For instance, there are no reports of agrammatism of the type associated with Broca's aphasia in DAT. There are also no reports of disproportionate difficulty with repetition; in fact, unhidden repetition in the form of echolalia and palilalia is one of the characteristics of late-stage DAT. And finally, there are no descriptions of phonologic disturbance; that is, DAT patients apparently do not violate the phonotactic constraints of their native language (using nonnative sounds or sound combinations) or make errors in prosodic aspects of language.\(^{3,9,12}\) We emerge with a picture of language breakdown in DAT that is quite specific: Semantic and pragmatic deficits are marked; morphosyntactic deficits are rare, and phonologic deficits are rarer still.

Second, many authors have attempted to describe the language disturbance of DAT by com-
parison with focal aphasia, arriving at the conclusion, for example, that transcortical sensory and Wernicke's aphasias are frequent in DAT. \(^{128}\) I have avoided classifying the language disturbance of DAT as an "aphasia" for two reasons. First, although the term aphasia can be used to describe any language disorder, it has become associated with syndromes where language disorder is primary, or significantly worse than any other symptom. This is not true for the DAT population. Although the language of DAT does manifest some typical aphasic symptoms (anomia, semantic paraphasias, and comprehension deficits), the language disorder of DAT is not primary or isolated but rather persists among other intellectual impairments. I concur with Holland et al that "aphasia...is a difficult, if not glib, diagnosis to apply in such cases. In the context of general deterioration of intellectual function, language disorder typically becomes intertwined with so many concomitant neurobehavioral changes as to render the term aphasia almost useless." \(^{127}\) Second, identifying the language disorder of DAT as an aphasia may imply (by association with focal aphasias) that there is a static quality to the disorder and that there is the possibility of recovery. However, because DAT is relentlessly progressive and degenerative, so are the language disorders associated with it. The language deficits are continually changing and recovery has never been observed. The language patterns of focal aphasias may be similar to the language disorder of DAT in some ways, but also differ in many important respects (see also Ulatowska et al for discussion). \(^{128}\) This must be remembered when planning research, considering diagnoses, or contemplating treatment.

Research: Dissociations and Associations

Four areas of current research on language functions in DAT will be reviewed here: semantic impairment, syntactic preservation, pragmatic deficits, and brain-behavior relationships.

SEMANTIC IMPAIRMENT IN DAT

Anomia (word-finding difficulty) is the earliest and most common language symptom of DAT. As such, accurate description of the symptoms and origins of anomia in DAT has been the subject of much research.

The earliest explanation for anomia in DAT was a visual perceptual deficit. Several pieces of evidence support this claim: naming by DAT patients is improved if the patients are allowed additional sensory cues (e.g., touching the object). \(^{7}\) DAT patients often use the name of an object that is visually similar to the stimulus in tasks of confrontation naming (e.g., calling an anchor a "hammer"). \(^{10}\) these patients are better able to name objects that are very familiar and thus require less visual recognition (e.g., body parts) than less familiar objects. \(^{18}\) Kirshner, Webb, and Kelly systematically investigated the effect of visual perception on naming by comparing object naming of actual objects, photographs, line drawings, and visually degraded drawings. \(^{79}\) Their results indicated that perceptual difficulty did play a significant role in anomia of DAT patients. However, problems with this theory are obvious: First, it does not explain why DAT patients are anomic in spontaneous conversation where no visual perceptual skills come into play, \(^{72}\) and, second, the theory is not consistent with the finding that most verbal paraphasias in DAT are substitutions of semantically (not visually) related object names. \(^{126,70}\) Further evidence against the visual perceptual argument comes from recent studies that demonstrate that DAT patients with significant anomia perform relatively well on independent measures of visual form discrimination. \(^{20,61}\)

The search for clues into the origin of anomia in DAT has continued beyond the visual perceptual theory and has focused on specifically semantic deficits, that is, problems in (1) impaired lexical access and (2) deterioration of lexical representations. There is some evidence that underlying lexical representations are intact and that naming difficulties arise from a problem in lexical access or retrieval for verbal production. Evidence supporting this view includes the findings that:

1. In tasks of confrontation naming, DAT patients can often give a related name or circumlocution, suggesting that they know much about the meaning of the word but cannot find the exact name (e.g., "cutter" for saw; "this is for your eyes" for glasses). \(^{11,70}\)
2. Comprehension of words is generally superior to production of the same words, indicating that the underlying representation can often be accessed in a passive comprehension task when the name cannot be generated or retrieved on demand. \(^{70}\)
3. DAT patients can utilize phonemic cues to help retrieve words, indicating again that the information is there but cannot be easily retrieved. \(^{52,30}\)
4. There have been several reports of DAT patients using gesture to indicate the function of an object that they could not name, suggesting that the deficit is limited to
lexical retrieval and may not affect basic symbolic representations presumed to underlie both gestural and language productions.19,30

Another source of support for this view comes from semantic priming data that indicate that subconscious semantic associations may be intact in DAT.98,99,100 The findings of semantic priming in DAT are perhaps the most curious of all and require some explanation. Several researchers have demonstrated that, like normal subjects, DAT patients react faster in lexical decision formats (i.e., is it a word?) if the target word (e.g., nurse) is preceded by a related word (e.g., doctor) than if it is preceded by an unrelated word (e.g., shoe).101

Initially, this was taken to indicate that underlying semantic associations were intact and that the anoma was not the result of permanent underlying semantic problems but must be an effect of impaired lexical access. However, findings from other studies have not always confirmed intact semantic priming in DAT. For instance, Smith et al found that DAT patients did not exhibit any effect of priming,126 and Albert and Milberg found semantic priming only in a subset of DAT patients.4 Several other studies have found that DAT patients actually show greater priming effects than control subjects.20,30 In addition, Chertkow et al found that hyperpriming in DAT was associated specifically with words that were shown to be semantically degraded on a variety of other tasks (e.g., responses to probe questions).20

In summary, the positive results of the early studies suggested intact semantic representations in DAT, but follow-up studies have demonstrated abnormal responses to semantic priming including (1) lack of priming effects; (2) subgroup differences; and (3) hyperpriming. Perhaps most interesting, the finding of hyperpriming appears to coincide with degraded semantic representations for particular words and has been used to support the theory that DAT patients suffer from disrupted semantic representations.20 It is to this possibility that we now turn.

Perhaps the first convincing evidence for the proposal that anoma in DAT is a reflection of impaired semantic representations was presented in a case study.49 Through a variety of methodologies to examine lexical knowledge (e.g., naming, verbal, and nonverbal match to sample), Schwartz et al were able to attribute the naming errors (e.g., calling a dog a "cat") to an erosion of referential boundaries. This finding has been supported by observations that many DAT naming errors are within-category errors (e.g., "truck" for car), a finding consistent with a deficit in the underlying conceptual and semantic representations.20 Additional evidence that representations within semantic categories may be degraded in DAT comes from studies that have demonstrated correspondence between naming and word comprehension errors and consistency of response across several testing sessions.20,54,71,89 These findings are consonant with the notion of central degeneration of lexical representation, which would be expected to permanently affect both comprehension and production of specific items. Although there is a growing consensus that DAT patients do exhibit evidence of impaired lexical representations, there are only general proposals about the type of disruption that occurs within the lexical representation. For instance, Schwartz et al discussed the problem as a "breakdown in the structure of the underlying categories,"102 while Chertkow et al referred to the "degraded" semantic representations,20 and Grober et al proposed that the representations are disorganized.10 Specific models of the exact nature of the representational deficit will undoubtedly be the subject of future research.

In summary, word-finding problems are among the earliest and most obvious symptoms of DAT, but the underlying cause(s) of anoma remain disputed.5,11,30 Although most researchers concur that the origins are primarily cognitive, there is still considerable disagreement about whether the deficit is best characterized as a processing limitation that interferes with lexical access or as a deficit in the underlying lexical representations. Further, because DAT patients have significant problems in attention and memory, correlations between performance on tests of attention and tests of naming have led other researchers to attribute anoma in DAT to problems in attention and concentration.103,110 This review of recent research suggests that the anoma of DAT is heterogeneous and that no single explanation can account for the naming performance of any group of DAT patients. Visual perception, attention, lexical access, and deterioration of lexical representations all play a role in anoma of DAT. It is likely that individual patients may be more or less affected by one particular deficit and that groups of DAT patients fall into distinct subgroups demonstrating anoma for different reasons.51

SYNTACTIC ABILITIES IN DAT

Although most descriptions of language in DAT have observed that syntactic ability appears intact, few experimental investigations have addressed this question. The earliest detailed investigation of this phenomenon was Whitaker's description of a severely demented patient who spontaneously corrected agrammatic but not semantically anomalous sentences in repetition (e.g., "There are two book on the table" was repeated as "There are few books on the table")
while "The book is very happy" was repeated verbatim. This finding was taken to indicate that grammatical competence was selectively preserved and, therefore, must be somehow autonomous from the rest of cognition. Schwartz, Marin, and Saffran also examined a single subject in detail and found that their patient, despite severely deteriorated semantic ability, was able to manipulate syntactic structures (e.g., turn an active in a passive), again supporting the notion that the syntactic processor may in fact be special and neuropsychologically insulated from cognitive and cortical degeneration seen in DAT. Subsequent attempts to address this issue have found DAT patients to produce syntactically complex and well-formed sentences in spontaneous speech as well. Kempler et al evaluated the spontaneous speech of 10 DAT patients and 10 normal controls, and demonstrated that the spontaneous speech of DAT patients contained few syntactic errors and many lexical semantic errors. More important, the range and frequency of sentence types (e.g., relative clauses, questions, and adverbial clauses) were almost identical to the normal controls (rank order correlation, $p = .9835; p < .0000$), and there was no difference between the DAT patients and the control population on a measure of structural complexity. A second study reported by Kempler et al further confirmed the relative preservation of syntactic competence throughout a dictation task adapted from Schwartz et al. Twenty DAT patients and 20 normal controls were asked to write short phrases containing a homophone, a word that has one pronunciation (s) but two spellings (sea/see) and two meanings (body of water/visual perception). In each case, the words can be disambiguated by semantic cues (a word with a similar meaning such as "lake-sea," "look-see") or a syntactic cue (a word with little semantic content but definite syntactic restrictions on what can follow such as "the sea," "I see"). The DAT patients made significantly more errors than did the control subjects with both types of cues, and significantly more errors with the semantic cues than with the syntactic cues, suggesting that the disease does seem to spare the ability to use syntactic (more than semantic) knowledge in a writing task of this sort.

Comprehension of syntax has not proven to be as consistently intact as production of syntax. Schwartz, Marin, and Saffran demonstrated intact comprehension of four syntactic forms (active, passive, preposition, and comparative adjectives) by a single demented patient. However, Emery, using the Test of Syntactic Complexity and the Chomsky Test of Syntax, has documented syntactic comprehension deficits in a larger sample of 20 DAT subjects. These tests evaluate the patient's comprehension of syntax by eliciting verbal responses to grammatically complex stimuli (e.g., "The dog was bitten by the cat; which animal bit the other and which was bitten?") or gestural responses to similarly complex auditory stimuli (e.g., "Mickey tells Donald to hop up and down; make him hop."). In contrast to Schwartz et al's findings of preserved comprehension of grammatically complex structures, Emery found significant impairment in DAT patients' ability to process syntactically complex grammatical constructions.

It is not simple to evaluate the intractness of grammatical knowledge with conflicting data: intact production versus impaired comprehension. Uneven profiles of grammatical abilities across tasks have been observed in other forms of brain damage, and explanations for these dissociations fall into two broad categories: those that postulate modality specific blockage (e.g., a motor output problem in the case of preserved comprehension and impaired production in Broca's aphasia) and those that postulate selectively impaired independent processors for different grammatical tasks. The dissociation between intact syntactic production and impaired comprehension in DAT may have another explanation: Overall memory and processing demands may have affected performance on comprehension tasks more than production tasks. Emery argued against this explanation, citing the finding that DAT patients were able to repeat all of the stimuli accurately even though they could not comprehend them, suggesting that the problem is not necessarily one of memory but more likely the result of deterioration of complex syntactic processing. A recent study by Smith investigated the relative contribution of syntactic knowledge and general processing demands to performance on tests of syntactic comprehension. Smith presented a group of 22 DAT patients with a picture-pointing task to assess comprehension of reciprocals and reflexives, systematically varying elements that contribute to processing difficulty and syntactic complexity (e.g., two- versus four-choice response arrays and different position of the reflexive in the sentence). The results demonstrated that both DAT patients and controls showed performance decrements as task difficulty increased, and both groups showed a similar pattern of errors. These results were interpreted to indicate that comprehension deficits in DAT may be attributed to general processing limitations and not to a specifically syntactic deficit.

We are left with the finding that despite severe lexical semantic deficits, DAT patients retain grammatical ability in (1) spontaneous utterance production; (2) tasks of sentence comprehension; and (3) writing to dictation. This dissociation between syntactic and semantic abilities has been explained by recourse to the notion of automatcity. Syntactic structure has the characteristics of a domain that could be run by an automa-
PRAGMATIC DEFICITS

Pragmatics, the study of language use in context, includes a large variety of language skills from turn-taking to appropriate topic introduction and overall discourse structure, all generally considered within the context of interpersonal interaction. In its broadest sense, pragmatics covers everything relevant to communication beyond sentence structure and linguistic semantics, often including extralinguistic features of facial expression and body language. In comparison with other language functions, we know least about pragmatic disturbance in DAT. Because very little research has been reported on the pragmatic abilities of DAT patients, much of what can be said about this area is anecdotal and in need of confirmation (see Chapter 8 for more detail on discourse and dementia).

SOME ASPECTS OF DISCOURSE ARE CLEARLY PRESERVED IN DAT THROUGH THE MILD AND MODERATE STAGES: DAT PATIENTS TAKE CONVERSATIONAL TURNS WHEN APPROPRIATE AND OFTEN PRODUCE SOCIAL AND VIRTUALIZED PARS OF THE CONVERSATIONS (E.G., GREETINGS AND LEAVE TAKINGS) WITH APPROPRIATE TIMING, AFFECT, AND LINGUISTIC STRUCTURE. THESE OBSERVATIONS INDICATE THAT DAT PATIENTS ARE ABLE TO ADHERE TO BASIC STRUCTURES AND OBEY PRAGMATIC RULES OF SOME VERBAL INTERACTIONS. HOWEVER, THERE ARE STILL PRAGMATIC PROBLEMS EARLY ON, SUCH AS A TENDENCY TO REPEAT THINGS UNNECESSARILY AND TO LOSE THE TOPIC OF THE CONVERSATION. AT THIS STAGE THE DEFICITS ARE OFTEN ATTRIBUTED TO FAILING ATTENTION AND MEMORY.

BY THE MILD STAGES OF THE DISEASE, THE DISCOURSE OF DAT PATIENTS OFTEN BECOMES IRRELEVANT, LACKING IN TOPIC COHESION, AND GROSSLY INSENSITIVE TO THE NEEDS OF THE LISTENER. Hutchinson and Jensen analyzed conversations of 10 dementia patients (and 5 healthy controls) and found several pragmatic abnormalities including: (1) patients produced fewer utterances and more turns, resulting in fewer utterances per turn; (2) patients produced more directives (utterances aimed at getting the listener to do something, such as explain or clarify); (3) patients produced more utterances for which no intent could be determined, and (4) patients initiated more new topics, often inappropriately.

Further investigations of discourse in DAT have confirmed and expanded Hutchinson and Jensen's original findings. Particular attention has focused on the production of decreased information content and lack of coherence. Ripich and Terrell suggested that the perception of conversational incoherence on the part of DAT patients might be attributed to "absent pieces of discourse that should provide relationships between preceding text and that which follows." Ulatorska et al found that decreased informativeness could be traced back, at least in part, to problems in reference such as the overuse of demonstratives (E.G., here, there) and ephoric reference (E.G., this without a clear antecedent).

Still unaddressed are the underlying causes of the discourse problems in DAT. These deficits could be secondary to existing and documented problems such as anemia, decreased attention, and poor memory. On the other hand, they may result from more general pragmatic deficits that impair the ability of the DAT patient to take the perspective of the listener and to judge what information is important in the particular discourse. Further, DAT may selectively affect specific discourse knowledge. For instance, certain discourses are structured largely on the basis of internalized scripts—rules about what type of information is
LANGUAGE CHANGES IN DEMENTIA OF THE ALZHEIMER TYPE

It is well known that language structures and other cognitive processes reflect a great deal of the underlying neural structure of the brain. For example, neurologists have observed that patients with neurological disorders often experience difficulties in their cognitive processes. This suggests that there is a strong connection between the brain and cognitive functions.

One investigation specifically looked for associations between discourse impairments and other language deficits. Nicholas et al. compared DAT patients' performance on the Boston Naming Test (BNT) with elements of empty speech in narrative descriptions of the cookie theft picture in an attempt to evaluate the claim that discourse incoherence could be attributed to anomia. The authors reported a significant negative correlation between the score of DAT patients on the BNT and the use of indefinite terms (e.g., "thing" and "stuff") and a significant positive correlation between the BNT and the production of "content elements" (i.e., references to characters and activities, in cookie theft stories). They concluded that the naming deficit did not underlie the emptiness of discourse, presumably because many other measures of discourse emptiness (e.g., paraphasias, pronouns with antecedents, and deictic terms) did not correlate with the BNT scores. The data, however, also suggest that at least some of the referential problems that make discourse difficult to follow might be a result of anomia. That is, to the degree to which patients are anomie and substitute or omit content elements, their discourse will be difficult to interpret. Nicholas et al. are undoubtedly correct in their general conclusion that the anomia does not underlie the discourse deficits, but it is unarguable that the anomia does contribute to the observed discourse deficits.

Further description of the discourse problems in DAT can be used to delineate which aspects of pragmatics are related to which aspects of linguistic and nonlinguistic cognition. For instance, it can be hypothesized that some aspects of discourse (e.g., topic maintenance) depend on recent memory but may be independent of lexical and morphosyntactic function. Therefore, we could predict that these features would correlate with decreased memory and be difficult early on in the disease. Conversely, some aspects of discourse are more structural (e.g., the use of definite and indefinite articles to signal new versus old topics) and may be retained with morphosyntactic abilities. Other aspects of pragmatics may be unrelated to either memory or morphosyntax (e.g., politeness and use of speech registers) and therefore should not correlate with either. At this time, these are only conjectures to be investigated in the future. In summary, although little research has been reported to date, DAT is a likely place to start identifying the cognitive bases of pragmatic knowledge.

**BRAIN-BEHAVIOR RELATIONSHIPS**

Any neurogenic language disorder provides the opportunity to learn more about the language, the brain, and the relationship between the two. By correlating language disturbance with various types and locations of brain lesions, we have learned a great deal about the neural representation (lateralization and localization) of language. Although the language and neuro-pathologic changes associated with DAT are not as focal and specific as in cases of cerebral vascular accident (CVA), neither are they global or diffuse. For instance, we have seen that language changes affect semantics and pragmatics preferentially but leave grammatical and phonologic abilities relatively spared. Likewise, the neurologic picture is multifocal: Excessive numbers of neurofibrillary tangles, plaques, neuronal loss, gliosis, and amyloid angiopathy are found in association cortex and in the limbic system, but the primary motor and sensory areas are characteristically spared.

There are both direct and indirect methods of investigating the neurologic bases of language deficits in DAT. Direct observation of neuropathology is possible only through brain biopsy or autopsy. Because of the time course of the disease, direct histologically derived clinicopathologic correlations are difficult to specify in DAT; the distribution of plaques and tangles at postmortem can tell you little about the relationship between the brain and language performance at the point of interest some years earlier. Nonetheless, grossly spared versus grossly pathologic areas of the brain correlate with what is known about behavior in DAT and contribute to our theories of brain function. Chui reviewed clinicopathologic correlations in DAT and associated the fluent speech production of DAT with sparing of the primary motor and sensory cortex, while the impaired semantics was associated with depo-sited temporo-parietal association cortex. Clinicopathologic correlations, then, suggest that expressive fluency and syntactic ability may be subserved by primary motor and sensory areas in the anterior portions of the cerebral hemispheres, while semantic abilities depend on temporal and parietal association cortex. By inference, this strong association between fundamental linguistic and basic sensorimotor abilities lends support to theories of brain function that
have stressed the functional and even evolutionary connections between neural control of motor and sequencing skills and the specialized linguistic capacity of man's left hemisphere. 71

Noninvasive, indirect measurements of brain structure and function have the distinct advantage of allowing comparison of brain structure and function at or near the time of language evaluation. In vivo measures can be divided into two general categories: those showing brain structure—computed tomography (CT) and magnetic resonance imaging (MRI)—and those showing brain function—electroencephalogram (EEG), positron emission tomography (PET), and single photon scanning (SPECT). Studies of structural abnormality in DAT have been relatively unrevealing. DAT patients tend to have enlarged ventricles and widened cortical sulci on CT and MRI, and these changes generally do not correlate with the presence or severity of dementia. 62,68.83 Although this is essentially a negative result, it highlights the interesting fact that structural abnormality on CT and MRI scans is in no way prerequisite to serious, permanent language disturbance.

Studies of brain function in DAT have shown that the structural damage that may be minimal or invisible on CT or MRI has significant consequences for brain function as measured by blood flow or glucose utilization. Decrease in cerebral blood flow is a consistent finding in DAT; 13 it appears to correlate with severity and to be most pronounced in the parietal regions. 63 The particular patterns of brain dysfunction in comparison with language deficits are of interest here. Recent research has demonstrated that there are subgroups of DAT patients that present with primarily language or constructional deficits, and these subgroups display different patterns of cortical hypometabolism in the expected directions: Patients with marked anomia demonstrated lowered metabolic rates in the left temporal regions; patients with focal visuo-constructive impairment showed relatively more hypofunction in the right temporal and parietal regions. 19,41,88 These studies extend our notions of brain-behavior relations to include measures of glucose utilization. To date, most studies appear to coincide with the familiar division of function between the hemispheres and between the lobes that we have come to accept from years of clinicopathologic correlations.

The next step toward deciphering the neurofunctional code is to perform detailed case studies in which we can correlate specific language and cognitive deficits with specific patterns of brain dysfunction. One case study with serial PET scans of a DAT patient has attempted to do this. 88 In this case, although the patient showed significant memory deficits early on, the initial PET scan showed no abnormality. In addition, although no additional neuropsychologic deficits appeared during the course of the study, the patient showed decreased parietal metabolism in the two later scans. This case highlights the lack of correlation between observed behavior and measurable brain function when DAT patients are examined individually.

In summary, the investigations into brain-behavior relationships with DAT tend to uphold traditional models of cerebral function of language, but force us to expand those models in certain ways. It is clear that gross left-hemisphere structural change is not prerequisite for a lasting language disorder and that measures of brain function (e.g., blood flow) are not yet sensitive enough to correlate with language dysfunction on an individual case basis. It is also clear that the type of structural and functional damage seen in DAT spares primary motor and sensory regions as well as areas thought to subserve syntactic function. Further research into brain function in DAT will undoubtedly add to our knowledge of the neural representation and neuropsychology of language.

Clinical Issues

Any clinical consideration of dementia or DAT must include a discussion of evaluation, differential diagnosis, and potential treatment. Each of these issues will be taken up in turn. For further discussion of diagnostic and treatment issues, see Chapters 15 and 15.

EVALUATION

The goal of the language evaluation is to reach or confirm a diagnosis and, ultimately, to develop a treatment plan. Language evaluation of dementia patients combines two elements not usually relevant to other populations: age and multiple cognitive deficits. The special considerations that need to be addressed because of the relatively old age of these patients are well described in a recent chapter by Groher, and include admonishments for the clinician to: (1) allow ample time for all aspects of the evaluation to compensate for the inevitable slowing that accompanies age; and (2) compensate to the degree possible for peripheral disturbance (e.g., decreased vision and hearing) within the test environment to prevent these factors from contaminating the evaluation of central language abilities. 31

The second important point to remember in evaluation of a demented patient is the likelihood of multiple cognitive deficits in addition to language disturbance. It is essential to establish the relative severity of nonlanguage deficits in memory,
attention, nonverbal problem solving, daily living skills, and spatial orientation. These factors will be important for reaching a diagnosis as well as for implementing a treatment program. For instance, severely impaired episodic memory (memory for personal experiences) will preclude direct teaching of any compensatory techniques (e.g., self-cuing); the self-care level will tell you what the patient's daily interactions are like and what type of language production and comprehension skills the patient needs in his particular social environment.

Both formal and informal tests should be used. Standardized aphasia batteries are valuable for their inclusion of many aspects of language functioning (naming, comprehension, reading, writing, spontaneous speech, repetition), and the Communication Abilities in Daily Living (CADL) provides additional information about functional skills. However, with the exception of The Arizona Battery for Communication Disorders of Dementia, standardized tests have little or no normative data for the demented population. Other specific tests (e.g., Token Test, Reading Comprehension Battery for Aphasia, and Boston Naming Test) are valuable for in-depth analysis of particular aspects of language. Analysis of language samples taken from conversation is also important because they provide a gauge of an individual's communicative abilities in a relatively unstructured situation.

**DIFFERENTIAL DIAGNOSIS**

Differential diagnosis refers to the characteristics that distinguish one disease from another. It is an important step in reaching a probable diagnosis and crucial in arriving at a useful treatment plan. With most dementias and DAT in particular, definite diagnosis can only be made on the basis of histopathology and is therefore limited to patients who agree to a brain biopsy or have come to postmortem. Therefore, there is a great need to improve our ability to diagnose DAT on the basis of clinical evaluation. The issue of the differential clinical diagnosis of DAT has been taken up by a special composite panel of NINCDS-ADRDA personnel that produced published guidelines for diagnosis of probable DAT. The criteria include documented dementia with deficits in two or more areas of cognition (including language), progressive worsening of cognitive deficits, no disturbance of consciousness, and the absence of other disorders or diseases that could account for the progressive deficit in memory and cognition. Some researchers have proposed somewhat more stringent criteria for the clinical diagnosis of DAT, requiring deficits in three areas of cognition and even suggesting that language disturbance be required for the diagnosis.

From the perspective of the speech-language pathologist, it is hoped that an adequate description of language in DAT will help to distinguish this from other forms of dementia, including multi-infarct dementia, normal pressure hydrocephalus, and dementia associated with extrapyramidal disorders. An adequate history of language symptoms and a thorough language evaluation should go a long way toward differentiating DAT from other forms of dementia. Language features that help to distinguish DAT from other dementia syndromes include: presence or absence of specific language symptoms, onset and course of language disorder, and order of appearance of language disorder vis-a-vis other problems such as motor disturbance. The remainder of this section will highlight the use of language disorders for discriminating between various dementia syndromes.

**Reversible Dementias**

There are many causes of potentially reversible dementia, including intracranial conditions (e.g., tumors and hydrocephalus), vitamin-deficiency states, endocrine disorders, drug intoxication, exposure to toxins or infections, and dementia resulting from affective disorders. Some studies estimate that up to 30 percent of the demented population may be suffering from a potentially reversible dementia. A thorough history and medical work-up are the best ways to differentially diagnose reversible dementias. A few of the more common reversible dementias, however, do distinguish themselves from DAT on the basis of language symptoms, or more specifically, their lack of language symptoms. For instance, the dementia associated with normal pressure hydrocephalus, resulting from impairment in the circulation of cerebrospinal fluid through the ventricles, is perhaps the most common potentially treatable cause of dementia. Hydrocephalic dementias generally present with a triad of dementia, ataxia of gait, and urinary incontinence. Although language deficits are generally not present, these patients may appear to have a sentence formulation problem or anoma caused by the substantial psychomotor retardation (mental slowing) that typically accompanies the syndrome. Unlike those with DAT, hydrocephalic patients can eventually generate the appropriate word or sentence without cues and have no language comprehension deficits. The absence of true language disturbance and the presence of a motoric (gait) disturbance early on in the course of the disease should alert the clinician to a clinical presentation inconsistent with DAT.

Another potentially reversible form of dementia is the "pseudodementia" associated with affect-
tive disorders such as depression. The dementia syndrome accompanying depression typically presents with psychomotor slowing, forgetfulness, disorientation, impaired attention, and disturbed ability to abstract and grasp the meaning of situations. Signs of cortical impairment such as language disturbance are not present. This dementia, if recognized and treated appropriately, has the same cure rate as any depressive illness.

**Subcortical Dementia**

Dementia is a common and debilitating symptom associated with extrapyramidal disorders such as Huntington's disease, Parkinson's disease, and progressive supranuclear palsy. Although motor disturbance is generally the earliest and most pervasive symptom in subcortical diseases, cognitive deficits occur in a large percentage of these patients. The standard symptoms of subcortical dementias include disorders of motivation, mood, attention, memory, personality, slowness of thought processes, and impaired ability to manipulate acquired knowledge (see Chapter 2). These patients generally do not present with agnosia or apraxia. It has also been assumed that, although communication is severely impaired by dysarthria and cognitive deficits, patients with subcortical dementias are not aphasic. However, recent research calls into question the status of preserved language in subcortical dementia. Demented patients with Huntington's disease are anomic and demonstrate a range of other language deficits including reduced syntactic complexity. Demented patients with progressive supranuclear palsy (PSP) have been noted to have deficits in word selection; and demented Parkinson's patients have been shown to have severely impaired syntactic comprehension. The data demonstrating impaired comprehension in Parkinson's disease are particularly important because they demonstrate a language deficit beyond language production, one that cannot be easily attributed to motoric or memory impairment. These studies that have found evidence of language deficits in subcortical dementia are consistent with data suggesting the presence of aphasia in patients with subcortical strokes and with an integrated view of brain systems in which damage to certain subcortical regions strongly influences the functioning of cortical areas traditionally tied to language ability.

**Cortical Dementias**

Alzheimer's and Pick's diseases are the two most common cortical dementias, but DAT is by far the most common, accounting for approximately 50 percent of dementias, and being 10 to 15 times more common than Pick's disease. They differ pathologically in that Pick's disease affects primarily the anterior temporal and frontal areas of the brain, while the pathology of DAT is concentrated more posteriorly, including parietal and temporal lobes and the hippocampi. Neuropsychologically, the two dementias are similar in preserved motor functions, impaired language, and altered personality through the early and moderate stages. They differ primarily in the areas of memory and visuo-spatial abilities that are spared in Pick's until later stages and impaired in DAT from the beginning. The descriptions of the language impairments in these two syndromes look grossly similar: anomaia in the early stages, progressing to more marked language disturbance with comprehension deficits in the moderate stages, progressing ultimately to echolalia and terminal muteness. A case study by Holland et al., in one of the few detailed descriptions of language dissolution in Pick's disease, documented a pattern uncharacteristic of language deficits in DAT. This patient presented with spoken language marked by literal paraphasias and some omissions of grammatical words, and eventually progressed to muteness. Auditory comprehension deficits were also observed. However, other language skills, including reading and writing, were relatively preserved and few other signs of dementia appeared through much of the 12½-year course. This case description is unlike DAT in several respects: The Pick's patient had preservation of memory and the "creative" aspects of language (semantics), while DAT patients typically present with significant memory deficits and semantic impairment. In addition, the Pick's patient appeared mute before the severe stages of the disease, but muteness is a late-appearing symptom in DAT.

Occasionally, DAT, Pick's, or Creutzfeldt-Jakob patients present with an isolated and progressive language disorder and only subsequently go on to develop other symptoms and an identifiable dementia syndrome. Another group of patients also present with progressive language deficits but do not develop a dementia syndrome. There has been lively discussion about whether or not these patients, if followed and tested carefully, will develop a range of neuropsychological deficits consistent with DAT or other dementia syndromes. So far, there has been little agreement about the appropriate diagnosis for these patients, and it has become clear that distinguishing the slowly progressive aphasia from dementia is not always easy. Serial evaluations documenting isolated and progressive language impairments can lead to a provisional diagnosis of slowly progressive aphasia. However, follow-up testing and thorough neuropsychological evaluations are essential in order to distinguish this syndrome from more widely recognized forms of dementia such as DAT and Pick's disease.
Mixed Dementias

The second most common cause of dementia is vascular or multi-infarct dementia (MID), which results from multiple cerebral infarctions and often presents with a combination of cortical and subcortical signs and symptoms. The symptoms are varied and are therefore sometimes difficult to distinguish from the symptoms of DAT. A thorough history can document abrupt onset, stepwise deterioration, and a fluctuating course that contrasts with the insidious onset and gradual course typical of DAT. Moreover, interim recovery of lost function is often observed in MID but not in DAT. A history of hypertension or strokes, with focal neurologic signs, also suggests vascular dementia.

One vascular dementia, the angular gyrus syndrome (AGS), resulting from posterior middle cerebral-artery lesions, has been particularly easy to confuse with DAT. The lesions in AGS are often too small to be visible on CT scans, and focal motor-sensory signs may be absent, leaving diagnosis largely dependent on neuropsychological findings, which appear similar to DAT, particularly in the area of language. AGS presents with fluent aphasia, alexia with agraphia, acausalia, left-right disorientation, finger agnosia, and constructional disturbances. The differences between AGS and DAT lie mainly in the preserved nonverbal memory and preserved awareness of language disorders in AGS.

Two important caveats must be made regarding the differential diagnosis of DAT. First, although up to 84 percent of dementia patients are clinically diagnosed with a primary degenerative dementia consistent with DAT, the diagnosis is complicated by factors of accuracy and specificity: 20 percent or more of cases with the clinical diagnosis of Alzheimer’s disease are found at autopsy to have other conditions and not Alzheimer’s disease, and dementia patients, like most older patients, tend to have multiple disease processes at work. For instance, in the case of DAT, 15 to 20 percent of the patients have been shown to have pathologic features consistent with both DAT and multi-infarct dementia. These facts force us to temper any generalizations we make about the behavior of clinically diagnosed DAT patients without histologic confirmation.

The second problem for differential diagnosis is variability of language symptoms within the DAT population. Alzheimer patients differ from one another. Some of the differences may be because of premorbid dispositions, and some may be because of stage of the disease. However, there remains a good likelihood that differences observed in language functions will be traced to subgroups of DAT patients with different etiologies. The most frequently mentioned subgroup of patients are those with genetic or familial DAT. The association between familial DAT and language disorders has been difficult to ascertain because researchers have found correlations between language deficits and positive family history of DAT, negative family histories, as well as no relationship between language disturbance and family history. Other subgroups based on language differences have been proposed, citing correlations between language disorders and age at onset and rate of progression, but these too have not been replicated. Therefore, it is clear that some DAT patients show more or less language disturbance than others, but the extent of these differences and the source of the variation are yet to be determined.

Treatment Implications

Once a diagnosis has been reached, a treatment strategy is designed. Although there is currently no cure for DAT, it is possible to compensate for some of the language problems described in this chapter. Candidacy for treatment will depend on the stage of the disease, the patient’s social environment, the patient’s awareness of his or her deficits, the nature and degree of other (e.g., cognitive) impairments, and, finally, the clinical judgment of whether treatment can help (see Chapter 15 for perspectives on treatment alternatives).

Traditional direct language therapy, as used to treat focal aphasias, might not be appropriate with DAT patients for several reasons. It is important to remember that DAT patients have a language disorder that differs in crucial ways from typical aphasias. For instance, unlike patients with focal aphasia, DAT patients present with intact grammatical and phonologic abilities, and the bulk of treatment must focus on semantic and pragmatic deficits. Because there has been a substantial body of research in the area of lexical semantics in DAT, there is now information that can be used to structure treatment. Unlike focal aphasia, where the anomia can be largely attributed to impaired lexical access, the anomia in DAT is probably polygenic (having multiple causes). Therefore, treatment and compensation will have to be multidimensional and tailored to individual patients. To the extent that visual perceptual impairment and lexical access deficits underlie the anomia, environmental support (e.g., good lighting) and lexical probing (Is it an X?) may alleviate the problem. However, because at least some of the anomia appears to stem from degraded lexical representations, probes, cues, and environmental support are less likely to be of any help. In these
instances, because research has demonstrated that superordinate category information remains relatively intact in DAT, communication might benefit from use of superordinate category names (e.g., furniture and tools) rather than basic level names (e.g., footstool and socket wrench).10

It is also crucial to bear in mind that the language disorders of DAT are embedded in a dementia syndrome and therefore occur among other, often more severe, cognitive deficits. These other aspects of the dementia preclude the use of traditional therapeutic techniques that rely largely on self-monitoring and on the acquisition of strategies to aid performance. Therefore, therapeutic intervention must emphasize compensation for lost function with little emphasis on learning. This includes indirect therapy such as counseling for the patient's caregivers and investigation of methods to enhance the patient's communicative abilities. A few compensation strategies that might prove useful with DAT patients are discussed below. Remember, however, that there is still very little research demonstrating positive effects of these approaches on communication.

Gestural communication has been used to compensate for language deficits in aphasia and might be considered as an augmentative strategy with DAT patients.13,24 In fact, the literature on naming impairment in DAT includes several observations that DAT patients spontaneously pantomime the use of objects that they are unable to name.12,10 Although this indicates that DAT patients might be able to use gesture to compensate for anomia, several studies have also documented a decline in DAT patients' ability to produce and understand pantomime, which parallels deficits in spoken language and indicates that compensation through gesture may be difficult for this group.70,81 It should be noted, however, that the research has not found a one-to-one correspondence between word loss and gesture loss. Therefore, even if the patient cannot produce or understand a specific word, he may still be able to make use of a related gesture and vice versa. No research has systematically explored the efficacy of gestural compensation in the treatment of DAT patients.

Because DAT impairs multiple aspects of cognition, and many of these nonlanguage deficits will impair communication, language therapy need not be limited to treating only the language deficits. For instance, deficits in memory and orientation may result in language comprehension problems and pragmatic deficits of poor topic maintenance and discourse incoherence. Improving these nonlinguistic cognitive abilities would likely have beneficial effects on communication. Perhaps the oldest and most common cognitive intervention for dementia is “reality orientation therapy.” This consists of providing orienting information to the patient with the goal of improving all aspects of functioning that rely on orientation (including daily communication and participation in therapies). Studies on the effectiveness of reality orientation therapy suggest that although this type of therapy may have a positive overall effect, the benefits are difficult to document objectively.8

Other cognitive intervention strategies for DAT patients are drawn from the literature on methods to improve and compensate for memory deficits in normal aging.126 One technique for improving recall involves structuring the input to aid memory. If language addressed to DAT patients could be structured so that it is encoded more completely and remembered more thoroughly, language comprehension and discourse abilities that depend on memory might improve. In one series of experiments, DAT patients were asked to remember a series of objects that were presented in different conditions (e.g., objects, objects with an instruction to tell the function of each one, and objects with a motoric instruction) and asked to recall the list of items in various conditions.6 One important finding is that the input condition appears to have different effects at different stages of the disease. For example, for mild DAT patients, the conditions with multiple input modalities (e.g., object and action) did not appear to affect performance significantly, suggesting that multiple modality input does not enhance memory at this stage of the disease. However, for moderate and severe DAT patients, verbal input alone was the optimal condition for free recall, sparking speculation that a “pure verbal presentation enables these patients to use verbal repetition to a greater extent than in the other conditions.”6 These initial findings suggest that multimodality input may be of little help to memory in the initial stages and may be a hindrance later on. Other memory-enhancing strategies such as “subject performed tasks” during encoding (e.g., lift the cup, put on the glove) appear to enhance memory at all stages of DAT,87 indicating that some interventions, particularly those that rely on motor acts, may be more effective for the population as a whole. Although many investigations have demonstrated effects of encoding manipulations on memory performance, the effects of such manipulations on language performance are unclear.

One research project has tested the effect of altered input directly on language comprehension. Young and Kennedy presented two patients with severe DAT with pragmatic or linguistic orienting cues prior to a simple picture-pointing comprehension task.18 The pragmatic cue consisted of an “alerting strategy” in which the examiner called the subject's name, made eye contact, and then gave the instruction (e.g., “Point to snow plow”).
The linguistic cue consisted of a semantic context as in "Show me something that clears the road in winter. Point to snow plow." One subject profited from both cue conditions, but more so from the simple pragmatic alerting cue. The other subject's performance did not improve with either cue. This study highlights both the potential effectiveness for improving language comprehension by a simple alerting cue prior to stimuli presentation and the importance of considering individual differences in any treatment program: What works for one DAT patient may not work for another.

There are many other techniques that may be effective in enhancing communication with DAT patients, although there is very little research to prove their efficacy. For instance, a reduced rate of speech has been shown to improve auditory comprehension in normal elderly, and may be effective with DAT patients as well.45 Ouslin and Santo-Pietro46 and Bayles and Kasnian47 recommend many simple and potentially helpful alterations in caregiver's speech that might enhance communication with DAT patients. These include: using short sentences; paraphrasing a sentence when repeating it; avoiding open-ended questions; being redundant; avoiding analogies; establishing eye-contact before addressing the patient; using pictures, objects, and gestures to enhance the message; and allowing time for processing.

Finally, it must be emphasized that, because there is no cure for DAT, and direct treatment is of unproven value with this population, caregivers are crucial participants in any treatment for DAT patients. The caregivers must be educated about the disease itself (etiology, course, and complications) and counseled about the patient's impairments and abilities. There are now many good, comprehensive books that can help to educate caregivers.45,49,60,69 Thorough language evaluations will help the caregivers understand the communicative strengths and limitations of the patient, and repeat evaluations can be a valuable method of monitoring the progress of the disease. It is the caregivers who will be primarily in charge of implementing the therapeutic strategy on a daily basis, and it is they who will ultimately benefit most from improved communication.

Future Research

There are many places where our research needs to be expanded, and I will make only a few suggestions here. There is ample evidence that there are individual differences in language abilities and deficits within the DAT population and that these differences are not random. It is crucial that variability of language impairments be documented, and if reliable subgroups exist, they must be adequately described. Much of the inconsistency in current research findings will ultimately be attributed to variability within the population rather than variability in experimental paradigms. Considering the extent of behavioral variability within the DAT population, it is essential that future treatment for this population be empirically derived and tailored to individual patients.

Over the past decade, there has been an almost blind devotion to studying the lexical deficits in DAT to the exclusion of other aspects of language. This makes sense in that anoma is one of earliest and most apparent deficits in the disease. However, we have produced little basic research on pragmatic and discourse deficits that are also early and apparent in DAT, and we have produced even less clinical research to document how and when intervention is most effective. Both of these areas deserve more attention in the next decade.

Collaboration across disciplines must also be increased. This may come in the form of collaboration with psychopharmacologists who are testing new drugs and need behavioral measures to test the effect of their medicines on cognitive deficits. It may come in the form of collaboration with epidemiologists attempting to sort out the behavioral markers of familial versus sporadic subtypes of DAT. It should certainly come in collaboration with caregivers and other clinical specialists (e.g., primary care physicians, nurses, and occupational therapists) who communicate daily with DAT patients.

References


