CASE STUDY

ACTA Vol. 9, No. 2, 2011, 193-208
NEUROPSYCHOLOGICA

Received: 11.02.2011
Accepted: 30.06.2011

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PROGRESSIVE LANGUAGE AND SPEECH DISTURBANCES IN TWO DIFFERENT TYPES OF DEMENTIA

Maria Pąchalska[1(A,B,C,D,E,F,G)], **Beata Łukaszewska**[2(A,B,D,E,F)]

[1] Chair of Neuropsychology, Andrzej Frycz-Modrzewski Cracow University, Cracow, Poland
[2] Institute of Psychology, University of Gdansk, Gdansk, Poland

SUMMARY

Background The specific course of aphasia and other language-related disturbances in dementia depends not only on the nature and course of the specific disease, but also on the patient's cognitive reserves and premorbid linguistic competence, and it may be also overshadowed by other cognitive and behavioral problems. The picture is further complicated by the fact that different neuropathological etiologies can give identical clinical symptoms, and conversely: one and the same disease acting on the cellular level can give diverse clinical manifestations, both between patients and over time in a single patient. In any case, however, the endpoint is invariably the loss of speech and language functions, followed by organic mutism.

Case descriptions: This article presents two clinical profiles from the most common forms: fronto-temporal dementia (FTD) and dementia of the Alzheimer type (DAT). A general description of the speech and language disturbances most often encountered in each of these illnesses is followed by a brief case study from the authors' own clinical material.

Conclusions: The present paper suggests that in different types of dementia the profiles of aphasia and language related disorders are very specific, but in the course of the disease they increasingly overlap. The complex relation between cognition and language is among the foremost theoretical problems raised by speech and language disturbances in different type of dementia. Microgenetic theory can be used to explain why neither separating thought from language nor reducing the one to the other is an adequate solution.

Key words: language disturbances, fronto-temporal dementia, Steele-Richardson-Olszewski syndrome, dementia of the Alzheimer type

INTRODUCTION

Most dementia patients exhibit some disturbances of speech and language, ranging from dysarthria through aphasia to organic mutism, though these are often overshadowed by other symptoms of mental deterioration. The symptoms are diverse and changeable, however, which makes it very difficult to generalize. Some patients display symptoms that can easily be classified into linguistic categories (phonology, semantics, syntax), while in other cases there are pragmatic problems in interpersonal communication, often interpreted in the context of behavioral or cognitive deficits (Pachalska et al. 2011). The disturbances in verbal communication are sometimes mild, sometimes severe; they may appear late in the course of the disease, or they may be the first presenting symptom of dementia (Kempler 1995). This diversity results from a complex calculus, whose main factors include the pathogenesis of the disease itself, the characteristics of the speech and language behavior of the affected individual prior to onset, and the cognitive reserves available to cope with and compensate for the loss of speech and language functions (Stern 2002). Moreover, by the end stages, as the disease progresses, most of the distinctions are gradually wiped out as the deterioration becomes increasingly global. The beginning point and initial course of each illness is thus relatively specific, but the end point (mutism and death) remains the same.

For a variety of reasons, speech and language has not been a major concern in the study and treatment of dementia, while for many years dementia-related speech and language disorders were not considered aphasia (Ross et al. 1990, Harciarek & Jodzio 2005). Nevertheless, the negative impact of these disturbances on the quality of life of demented individuals is considerable, even if other problems are given more attention. The cognitive and behavioral deficits that define dementia for most people result from the atrophy of brain tissue and a consequent decline in mental efficiency. The course of dementia symptoms (i.e. the speed of decline, and the appearance of particular cognitive, affective, and behavioral symptoms in a particular order) depends in each case on the nature of the neural degeneration caused by the basic pathomechanism, the point at which the degenerative process begins, and the general direction in which it spreads, not to mention the patient's premorbid intelligence, habits, and predispositions. At the cellular level, each disease is accompanied by pathognomonic neuropathological features, but the clinical picture *in vivo* is seldom clear-cut, especially since neuroimaging techniques rarely give an unequivocal answer to the basic nosological questions (see Hodges et al., this issue). Though the search for non-invasive tests based on biological markers continues (Leszek et al. 2003), at present a definitive diagnosis of almost all dementive illnesses is only made possible by neuropathological results showing the characteristic cellular features of a given disease. The clinical diagnosis is always at best a reasonable inference awaiting neuropathological confirmation.

There do exist, however, identifiable neuropsychological syndromes of dementia that have been found to correlate at least roughly with particular dementive illnesses. For example, "Dementia of the Alzheimer's type" (DAT) is characterized by the insidious onset of problems with episodic memory appearing after age 60-65 (Mendez & Cummings 2003), often followed by depressive symptoms and confusion, then gradual loss of other memory domains and a general cognitive decline, with language disturbances and other neurological signs appearing relatively late. A diagnosis of DAT give roughly 75% confidence that the patient actually has the neuropathological features of Alzheimer's disease. Thus a neuropsychological diagnosis based on test results describes a clinical syndrome of dementia bearing certain characteristic features, not unrelated to the ultimate neuropathological diagnosis, but in an important sense independent of it. For example, a post mortem neuropathological finding that the patient did not have the amyloid plaques and neurofibrillary tangles characteristic of AD does not mean that the neuropsychological diagnosis of DAT was in error, but only that the neuropathology underlying the observed symptoms proved to be something other than AD.

In order to keep the discussion here within reasonable limits, only two dementive syndromes will be described here:

- Fronto-temporal dementia (FTD);
- Dementia of the Alzheimer's type (DAT).

Each of these descriptions will be supported by a brief case study, drawn from the author's own clinical material.

FRONTO-TEMPORAL DEMENTIA (FTD)

In fronto-temporal dementia (FTD), personality changes (consistent with the well-known picture of "frontal syndrome") usually play a much more important role than specific cognitive deficits (Pasquier & Delacourte 1998, Mendez & Cummings 2003, Kertesz et al. 2007; Pachalska et al. 2011). The diagnosis of FTD is most often based on the Lund and Manchester clinical criteria (Lund and Manchester Groups 1994), combined with a neuroradiological finding of frontotemporal atrophy consistent with corticobasal degeneration (Miller et al. 1998, Varma et al. 2002, Boccardi et al. 2002, 2003). Among the most important diagnostic criteria listed by the Lund and Manchester study (Gustafson et al. 2004) are the following:

- insidious onset with slow progression;
- early loss of insight;
- early signs of disinhibition and impulsivity;
- loss of mental flexibility (inability to adjust to changes of plans or circumstances);
- stereotyped, perseverative behavior (motor and verbal);
- utilization behavior (a tendency to pick up and manipulate all objects lying within arm's reach);

- hyperorality (frequently manifested in compulsive eating and drinking);
- difficult in staying focused on the task at hand.

Nevertheless, every one of these criteria has been challenged in the literature, especially since the difference between the clinical picture of FTD and that of DAT or other dementias may be more the timing (i.e. when the given symptom appears) than the specific symptomatology (Pachalska et al. 2011). Thus the disputes between "lumpers" and "splitters" rage on to the present (Kertesz 2005). For the present purposes, however, it is not essential to sort out the "alphabet soup" of proposed nosological entities that may or may not fall within the general category of FTD. It is surely safe to say that, since the neuronal loss associated with fronto-temporal lobar degeneration and its many variants causing FTD often occurs in areas critical for speech and language, aphasia symptoms tend to appear earlier in the frontal-temporal dementias than in DAT, and not infrequently constitute the first presenting symptom (Knopman et al. 1990, Snowden & Neary 1993, Neary et al. 1998).

In recent years the term "Pick's complex" (Kertesz 1998a, Kertesz et al. 2007, 2008; 2010; Ioannides et al. 2005) has come into use to denote the significant clinical and pathological overlapping between primary progressive aphasia, frontal lobe dementia and corticobasal degeneration. Kertesz (2005; for review see also Kertesz, this issue) argues that despite the diversity of the clinical picture, FTD is a clinical syndrome, within which one can further specify variants, including a behavioral variety with features of frontal syndrome, progressive aphasia, semantic dementia, corticobasal degeneration and progressive supranuclear palsy (PNP, also known as Steele-Richardson-Olszewski syndrome).

Aphasia in FTD typically begins with the diminished verbal spontaneity, demonstrated by diminished verbal fluency in neuropsychological testing, particularly when the task involves listing words that begin with the same letter (Kertesz 2003). There are frequent stereotyped comments, not always relevant to the context in which they are used (see Code 1997 for a comparison to the repetition of stock phrases in popular music). The combination of palilalia (repeating the same word over and over), echolalia (repeating the conversation partner's last words before beginning one's own utterance, cf. Lebrun 2003), late mutism and amimia (loss of spontaneous mimicry), known as the PEMA syndrome (Guiraud 1956), is typical of FTD; logoclonia, however (the tendency to overuse a particular word, substituting it for a number of different target words), is rare. A particularly salient feature of aphasia in FTD is difficulty in understanding sentence structure (Grossman et al. 2005).

In some FTD patients, the development of dementia symptoms other than language-related may be delayed, sometimes for many years, in which case we speak of "Primary Progressive Aphasia" (PPA), though the term itself and the relation of this syndrome to FTD remain controversial (Knibb & Hodges 2005, Selnes & Harciarek 2005). Already in the 19th century Arnold Pick had pointed out that progressive atrophy in the frontal and temporal lobes evoked a constellation of symptoms that included personality change and dysphasia (Kertesz

1998b, Selnes & Harciarek 2005), but Mesulam (1982**;** for review see also Harciarek & Kertesz, 2011) was the first modern author to demonstrate the existence of a syndrome of aphasia developing slowly in the absence of other symptoms of dementia or neurological signs. Luzzatti and Poeck (1991), Kempler (1995), and Mesulam (2001) thus identify the most common clinical features of PPA:

- a non-fluent aphasia with hesitant, dysprosodic speech, phonemic paraphasias, and impaired syntax;
- poor repetition;
- generally preserved comprehension;
- normal or near-normal performance on tests of other cognitive functions.

Another variant of FTD that has a noticeable and early effect on speech and language is semantic dementia (SD; see Lambon Ralph et al., this issue). Hodges et al. (1992, cf. Garrard & Hodges 1999) described the insidious onset and progressive breakdown of semantic knowledge, which affects not only language, but also memory, general knowledge, and object recognition. The result is usually a fluent aphasia, involving anomia with circumlocution and semantic paraphasia, single-word comprehension deficits, and reduced category fluency, typically accompanied by visual agnosia and other symptoms of a generalized inability to abstract conceptual meaning from concrete signs and symbols (Knibb & Hodges 2005; Kertesz et al. 2010).

Case study: ZE

This patient, a right-handed Polish female, was 72 years old when the first symptoms appeared. ZE was a prominent professor of medicine, a cardiosurgeon of international stature in her field of specialization, who spoke several languages fluently enough to lecture around the world. She had always been remarkable not only for her high professional and scientific qualifications, but also for her warm and outgoing personality. Her home had always been a international “salon” of sorts, and her dinners for specially invited guests were once legendary in Cracow. She had always enjoyed remarkably good health until the spring of 1999, when she noticed that she was more and more often forgetting daily tasks and leaving personal objects in unlikely places. Since her mother had died of Alzheimer’s disease several years earlier, ZE was alarmed by this and reported it to her family, but no one felt that the problems were sufficiently severe to merit attention. Later, she began to have sudden, unmotivated (and highly uncharacteristic) outbursts of rage, but again, her friends and family interpreted these incidents as the result of stress and fatigue associated with her busy professional calendar (which despite her nominal retirement would have exhausted someone half her age). She herself attributed her emotional lability to her latent fear of AD.

The first neurological symptoms appeared in late 2001, when her gait slowed and became ataxic, so that she had several falls (early falls are considered a sign of progressive supranuclear palsy, or PSP, a form of FTD; cf. Litvan et al. 1996, Kertesz 2003). Since the earlier behavioral symptoms were not taken seriously, and ZE developed, in addition to the motor symptoms, a Parkinsonian “mask,”

she was initially diagnosed with PD. However, dopaminergic treatment proved completely ineffective and even counter-productive (another sign of PSP). The final piece of the puzzle fell into place in May of 2002, when during neuropsychological testing the first author observed that EZ was unable to look upward or downward without moving her head ("vertical ophthalmoplegia" is virtually pathognomonic for PSP). However, her MMSE score in the first examination was normal (29/30 points). Neuropsychological testing (see also Pachalska et al. 2003) showed:

- progressive subcortical dementia; the severity of dementia, evaluated using the Clinical Dementia Rating (CDR: Hughes et al, 1982), was very mild in the first examination (CDR= 0.5), and mild in the second examination (CDR= 1)].
- reduced criticism, impaired working memory, dysexecutive syndrome (especially difficulty in planning activities and constructional apraxia),
- verbal and motor perseverations;
- limb apraxia;
- progressive behavioral changes. The severity of behavioral changes, evaluated using the Frontal Behavioral Inventory (FBInv; Kertesz et al. 1997, Polish version Pachalska & MacQueen 1998), was 38 of 72 possible points, much above the cutoff point that justifies a diagnosis of FTD (see: Kertesz et al. 2000; Pachalska et al. 2011). In the second examination the FBInv score was much higher (61 points).

The speech and language disturbances presented by ZE (with the exception of slowed speech tempo and slight, barely noticeable slurring, consistent with hypokinetic dysarthria, cf. Darley et al. 1975; Pachalska 2008), accompanied by dysphagia, appeared considerably later than the motor symptoms. There was also dysgraphia, manifested in distortion of letter shapes and difficulty keeping the written text on the lines of the paper. Later she began to make orthographical errors. The progress of dysgraphia is illustrated by Fig. 1.

Until very late in the progress of the disease ZE had no pathological scores on any aphasia tests, though it was noticed that, in comparison to her premorbid speech habits, her lexicon was distinctly impoverished. She became markedly less spontaneous, often smiling and nodding instead of speaking, even in response to a direct question. She seemed at that point to be moving in the direction of Luria's "dynamic aphasia" (Esmonde et al. 1996, Luria 1977), characterized by markedly diminished verbal output with relatively few patent errors.

One of the most salient features of ZE's speech at this stage was an increasing tendency to fall into various forms of pathological repetition, a common feature of both PSP (Lebrun et al. 1986) and Parkinson's disease (Benke et al. 2000). Initially, ZE in conversation often repeated the question she had just been asked before answering it, as though wanting to be sure that she had understood it correctly. For example:

Examiner: What time is it?
ZE: What time is it? Oh, ten o'clock.

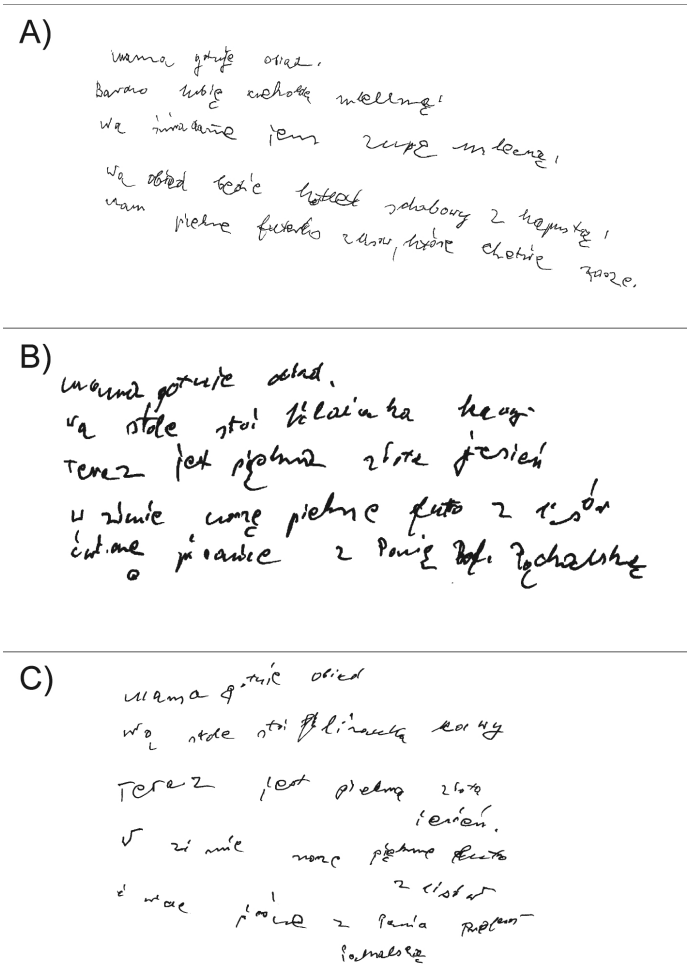

Fig. 1. Writing samples from patient ZE in October (A), November (B) and December (C) of 2002. The prompt text was identical in each case

Over time this habit developed into echolalia, and the repetition of the interlocutor's speech was less and less often motivated or followed by an appropriate response, as follows:

Examiner: It's very cold today.
ZE: It's very cold today. Yes, it's very cold.

The Western Aphasia Battery (WAB) showed the occurrence of aphasia, with an Aphasia Quotient (AQ) of 89.0, which was slightly lower one year later (84.0). The index of the Cortical Quotients (CQ) was on the borderline of norms, but was markedly lower one year later, with a CQ of 71.0. This was probably due to the fact that picture recognition scores are included in the CQ, and she did not like the task.

She also developed palilalia at this time, first involving the repetition of words, then syllables, then single phonemes, rendering it difficult to determine a sharp boundary between palilalia and stuttering (Lebrun 2003; Pachalska 2008). Unlike typical stuttering, however, she most often repeated the last syllable or phoneme of a word she was trying to use, rather than the first.

It is of particular interest to note that until a relatively late stage in her illness ZE was not only able to converse in English, but in fact showed less dysarthria and echolalia in this language than in her native Polish. She enjoyed speaking English and often spontaneously shifted languages during testing. However, as the disease progressed she fell into a global aphasia and ultimately ceased to display any verbal behavior at all.

DEMENTIA OF ALZHEIMER'S TYPE (DAT)

Though there are several different schemes of staging in DAT, for the present purposes it is most convenient to speak simply of early, middle, and late phases (Gustafson et al. 2004). The early phase is usually pre-clinical, with loss of short-term memory attended by confusion, which becomes increasingly troublesome until it finally prompts the patient (or the family) to seek medical attention. The late phase is a virtually complete loss of logical contact, accompanied by increasingly severe neurological disorders and terminating in death. What we are calling the "middle phase," then, is not so much a clearly delineated stage as a series of transitions that lead from the preclinical to the late stage.

Aphasia usually appears relatively late in the course of DAT. In the classical presentation, speech remains largely unaffected until the transition from the middle stage to the late stage, when aphasia joins other neurological symptoms, including motor problems. However, even in early DAT, although ordinary conversation may seem unaffected, patients often have more word-finding problems than do healthy elderly, and typically show below-normal scores in verbal fluency (Cerhan et al. 2002, Harciarek & Jodzio 2005). In neuropsychological testing, the task of naming as many animals as possible in one minute is usually easier for DAT patients than the task of listing words that begin with a certain letter, and shifting from the one task to the other almost always produces perseveration of the first task into the second (Pachalska 2008).

The steady cognitive decline of the middle-stage AD patient is accompanied by impoverished vocabulary, leading to increasingly severe word-finding problems and ending in the organic mutism of the late stage. Automatisms and metalinguistic comments occupy an increasing amount of the patient's discourse. At the same, the mental confusion typical of AD at this stage leads to apparently nonsensical utterances that begin to take on the features of aphasic jargon (Mendez & Cummings 2003). Some authors (Gates et al. 2002) have compared the typical picture at this stage to transcortical sensory aphasia. Logoclonia is a characteristic feature of the speech of DAT patients.

Case study: JG

This patient, a right-handed Polish female, age 67 at first contact, was referred for neuropsychological testing by her family, whose concern was aroused when on several different occasions she seemed to become completely lost in her own neighborhood, unable to find her way home when she was only a block away from her home of 20 years. Since she had recently experienced the loss of her younger brother, the family at first attributed the problems to emotional stress, and indeed the first diagnostic task (often a difficult one in early DAT, see Dierckx et al. 2004) was to differentiate between depression and dementia. The possibility that JG was suffering from depression rather than a neurodegenerative disease was made more likely by the fact that MMSE score in the first examination was at the low end of the range indicating mild cognitive impairment (MCI, 24 of a possible 30 points).

On her first attempt to draw a clock face (a widely used screening test for dementia) she drew a circle with marks at 12:00, 3:00, 6:00 and 9:00, but then the examiner noticed that she was looking at a clock on the kitchen counter (she was first examined at her home). When the clock test was repeated 30 minutes later and the clock was removed from her line of sight, she drew the clock face correctly, but when asked to show “3:05”, she drew four different hands (none in the right place, then expressed frustration that she “couldn’t make it come out right” (see Fig. 2A).

Her conversation seemed normal and her mood actually improved considerably during the first examination session.

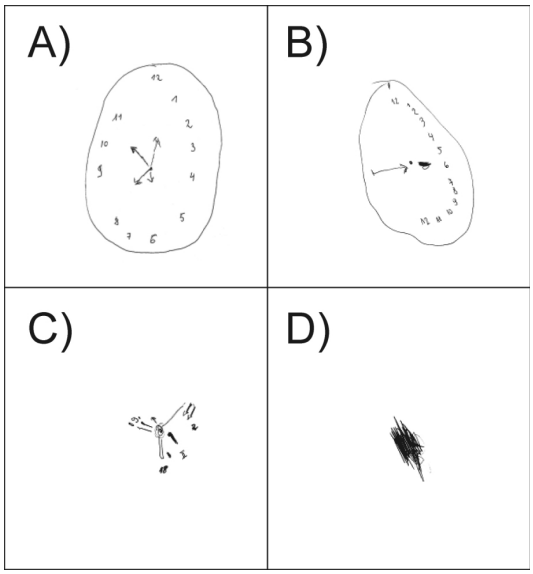

Fig. 2. Clock faces drawn by JG over the course of her illness. A) first examination: May 2005. B) June 2005. C) August 2005. D) September 2005 (one month before her death)

The Western Aphasia Battery (WAB) showed that the Aphasia Quotient (AQ) was on the borderline of normal at this point in time. One year later the AQ was slightly lower (89.0). More surprising was the Cortical Quotient (CQ) of 91.0, which was markedly lower a year later with a CQ of 65.0. This was probably due to the fact that picture recognition scores are included in the CQ, and this function is frequently impaired in DAT.

On the Boston Naming Test (Kaplan et al. 1983) she showed a sub-normal score (45 of 60 items properly named), while priming and prompting did not help her recover the names of objects she could not remember (see also Olszewski et al., this issue). In some cases, she indicated by periphrasis or gesture that she knew what the object was but could not think of its name. However, she called many of the animals simply "animal," and all the vegetables or fruits were "onions" (the first vegetable encountered on the Polish version of the BNT). When her attention was called to this after the test, she complained that she could not see (when later examined by an ophthalmologist, no abnormalities were found apart from a mild myopia, corrected by eyeglasses). On the test of verbal fluency, she did not reach norms in either task (category or initial letter), but her performance in the "words that begin with P" task was much worse (cf. Henry et al. 2004). Significantly, she fretted that she could not think of the names of any animals (the first, "category" task) that begin with P, and even after being reminded several times that the words did not have to be the names of animals, she clung to this restriction and could think of only two, which she repeated over and over.

When re-examined a month later, her mood was generally improved, and she remembered the examiners from the first meeting. However, her performance on the MMSE was now much lower (19/30 points), and all cognitive parameters were likewise reduced; her clock drawing was noticeably disturbed (see Fig. 2B). For the present purposes, however, most important is the fact that, although her score on the Frenchay Aphasia Screening Test was still within the normal range, conversation had become more difficult. She often left sentences unfinished or expressed herself so vaguely that it was not always possible to ascertain what she meant to say. The family likewise complained that it was becoming increasingly difficult to follow her meaning. When questioned as to her meaning she became flustered and occasionally expressed annoyance, sometime at her own inability to say what she wanted to say, sometimes at the obtuseness of the interlocutor, who in her opinion ought to know what she meant.

By the next monthly examination, the score on the Frenchay Aphasia Screening Test was now in the subnormal range (the task of identifying items on the "bridge" picture from that test now seemed entirely beyond her capabilities). There was now slight dysarthria (slurring), as well as logoclonia; during the Boston Naming Test almost half the prompts were now called "onions," and the word "onion" recurred in her conversation as well. Her mood was generally good, but she was often confused and was unable to stay with even simple tasks, though she tried hard to concentrate and was eager to please. Her writing was more agrammatic than her speech, with letter distortions and a general inability to maintain an even

line. Her clock drawing now bore little resemblance to a clock (see Fig. 2C).

By six months after the first examination, her speech was nearly incomprehensible (cf. Ball et al. 2004). Logical contact was fleeting and most standard neuropsychological tests had become difficult or impossible to administer. When asked to draw a clock, she made only a series of strokes, which she did not stop making until the paper was taken from her (see Fig. 2D). By eight months she required hospitalization, and after six weeks of steady deterioration she died. A post mortem neuropathological examination found the amyloid plaques and neurofibrillary tangles characteristic of Alzheimer's disease.

The relatively rapid progress of the disease in this case, dramatically documented by Fig. 2, is unusual for AD and requires some comment. Clinicians have long noted that AD tends to have a more severe course in persons of above-average intelligence (JG was a teacher, known before her illness for her wit and tendency to take a lively interest in things). It is now suggested by some authors (Stern 2002, Stern et al. 2003) that the reason for this counterintuitive phenomenon is that these patients, due to their high level of cognitive reserves, often do not show any clinical symptoms of illness until the disease progress is well advanced.

DISCUSSION

In a modular approach to brain work (Jackendoff 2000), one assumes that, somewhere in the brain, there is a thought module, which is both the origin of data presented to the language module to be transformed into a speech act and the destination of information decoded by the process of comprehension. In such a view, the language module (whether it is conceived as a single module or a set of modules forming a processing unit, cf. Jackendoff 2000) receives input in the form of thoughts, concepts, images, or the like, and produces a speech act as output, or, in reverse, receives phonemes or graphemes and transforms them through the application of linguistic rules into cognitions.

Postulating the existence of a "thinking module" at the apex of brain functioning seems reasonable enough, virtually instinctive, but it is attended by some serious problems, not the least of which is the difficulty in establishing where in the brain such a "thinking module" would be located. Could thinking be one of the executive functions performed by the frontal lobes, or is it a kind of meta-cognition situated in the strategic TPO region? Or are there two thinking modules, one anterior and one posterior? If so, how are they related to each other? The problem becomes more difficult the longer it is pursued. Perhaps it would be more expedient to question the necessity to postulate a distinct thought module, as Luria did (1977). There is really no such thing as "thinking," one might argue. Mentation would then be reduced to inner speech, i.e. a speech act that has not been realized through the motor segment (Brown 2004), or to imagination, assuming that the "stuff" of thought consists of words and pictures. This alternative has the further advantage of avoiding the "homunculus" fallacy, which necessarily emerges whenever a thought module is postulated.

Attempts to tease thought processes away from language-related processes begin by searching for a kind of "pure" aphasia (or dyslexia, or dysgraphia), in which thought processes are intact but specific language functions are disturbed. However, the incessant disputes in clinical psychology over the problem of defining and measuring intelligence should make us cautious about naively assuming that we know how to assess the quality of thinking in a manner that is not in some way dependent on language. In clinical practice, it is possible to be sure that there is intact thinking with disturbed speech only when the disorder is rather obviously peripheral in nature, sensory or motor, as in a hearing defect or dysarthria, i.e. affecting the receptor or effector responsible for receiving sensory signals or executing motor programs. The more central the point of disturbance, however, the more difficult it becomes to disentangle language from thinking. This is difficult to explain in the light of a modular system, in which the language module is conceived as a center for encoding and decoding, mediating between the inner world of thought and the outer world of behavior. If the language module is content neutral, as much of contemporary linguistics seems to assume, then it is only a messenger between "inner" and "outer" domains of information; if it is not content neutral, then the problem of distinguishing language from mentation becomes much more difficult.

It seems an obvious conclusion that the basic speech and language problem in dementia (as in psychosis) is that there is disordered speech because there is disordered thinking. Problems arise, however, when these concepts are tested critically in the clinic. A strictly psychometric approach can sometimes give us the illusion that mentation can collapse while leaving speech and language intact, or vice versa, but a careful observation of the patient's behavior very often gives the lie to this conclusion. In what sense can a nonsensical sentence be linguistically correct? For analogous reasons, semantics and syntax may seem to be separate processors within the language module, but when we examine what the brain actually does in real time to take in a sentence (MacQueen 2003), it turns out to be inordinately difficult to find "purely" syntactical or "purely" semantic operations (Pachalska & MacQueen, 2002).

The problem raised here is far too complex and far-reaching to solve here and now, though indeed dementia is a good point to begin the discussion, precisely because we are forced to ask the basic question: what is thinking? The very derivation of the word dementia (in past centuries a very general term for mental illness) points to "losing [one's] mind," which is echoed even in ordinary speech. The fact that we have narrowed the definition considerably and have a generally more "enlightened" attitude towards the problem should not disguise the fact that the patient ordinarily experiences the progress of dementia as rather literally a loss of mind. Moreover (again, setting aside for the moment our scientific predispositions), it is hard to imagine that one could lose one's mind (whatever we understand that to mean) without that fact finding some reflection in speech and language – and vice versa.

In microgenetic theory (Brown 1988, 2005) the process of forming a speech act is understood as a continuous stream, leading from drive through motivation to behavior, where specific mental functions are not "processors" in a data-processing system, but rather moments when the diversion or interruption of the flow produces particular consequences. The process of formulating a thought flows into the process of formulating a speech act in inner speech (Brown 2004), which in turn flows into a realized speech act. The mental process that leads to comprehension is a parallel flow, with inner speech mediating between rules and specific mental states. Just as a mutation and a change in the environment may cause the tree of evolution to branch at a given moment, so changes in circumstances or context, both inner (within the brain) and outer (in the world "outside"), cause the microevolution of a mental state to branch in a particular way. In such a system, nothing is entirely predetermined, but then again, nothing is entirely random. Brain damage, then, including neuron loss resulting from a degenerative process, affects the ensuing mental state(s) much as the damage to a branch affects the growth of the tree. Further growth (that is, the appearance of further stages in the affected process) may be stopped, but more often growth continues downstream from the point of damage, though its direction and force are likely to be changed. The nature of the symptom is not merely an absence, a failure to perform, defective output from a damaged function, but a vital clue to the nature of the process itself (Brown & Pachalska 2003). Involution reveals the process by which evolution works (Pachalska et al. 2011).

In microgenetic theory, then, the difference between disturbances of thinking and disturbances of speech and language is as much or more a matter of "when" as "where." The closer the damage is to the surface, the more its effects are felt in the fine articulation of the affected behavior, the last stages of microgenesis, and conversely: the deeper the damage, the greater the number of functions that are disturbed at the outset.

There is, of course, no cure for dementia, as there is no cure for the various diseases that cause it. It is possible, however, to slow its progress and mitigate its effects. In many cases, too, it may well be possible to prevent the involutionary processes from beginning. There is some evidence, though controversial, that regular mental and physical stimulation throughout life may prevent neural degeneration from beginning, even when there is a genetic predisposition (Stern 2002).

CONCLUSION

The present paper suggests that in different types of dementia the profiles of aphasia and language related disorders are very specific, but in the course of the disease they increasingly overlap. The complex relation between cognition and language is among the foremost theoretical problems raised by speech and language disturbances in different type of dementia. Microgenetic theory can be used to explain why neither separating thought from language nor reducing the one to the other is an adequate solution.

In our opinion, one of the great challenges facing neuropsychology in the century to come, as the world population ages (especially in Europe and the United States), is to develop therapeutic and preventive interventions that can keep a larger percentage of the world's population from "losing their minds" before their natural lifespan is over. This may require some discussion and debate about where and how research resources are allocated, especially since at present the involvement of neuropsychology with dementia has been almost exclusively in the area of diagnosis.

REFERENCES

Ball, M.J., Code, C., Tree, J., Dawe, K. & Kay, J. (2004). Phonetic and phonological analysis of progressive speech degeneration: a case study. *Clinical Linguistics and Phonetics, 18(6-8)*, 447-62.

Benke, T., Hohenstein, C., Poewe, W. & Butterworth, B. (2000). Repetitive speech phenomena in Parkinson's disease. J*ournal of Neurology, Neurosurgery and Psychiatry, 69(3)*, 319-24.

Boccardi, M., Laakso, M.P., Bresciani, L., Galluzzi, S., Geroldi, C., Beltramello, A. et al. (2003). The MRI pattern of frontal and temporal brain atrophy in fronto-temporal dementia. *Neurobiology of Aging, 24*, 95–103.

Boccardi, M., Pennanen, C.M., Laakso, M.P., Testa, C., Geroldi, C., Soininen, H. et al. (2002). Amygdaloid atrophy in frontotemporal dementia and Alzheimer's disease. *Neuroscience Letters, 335*, 139–143.

Brown, J.W. (1988). *The life of the mind. Selected papers*. Hillsdale, New Jersey, USA: Lawrence Erlbaum Associates.

Brown, J.W. (2004). Word concept, verbal hallucination, and inner speech in neuropsychology. *Acta Neuropsychologica, 2(2)*, 111-129.

Brown, J.W. (2005). *Process and the authentic life: toward a psychology of value*. Frankfurt & Lancaster: Ontos Verlag.

Brown, J.W. & Pachalska, M. (2003) The nature of the symptom and its significance for neuropsychology. *Acta Neuropsychologica, 1(1)*, 1-11.

Cerhan, J.H., Ivnik R.J., Smith G.E., Tangalos E.C., Petersen, R.C., Boeve B.F. (2002) Diagnostic utility of letter fluency, category fluency, and fluency difference scores in Alzheimer's disease. Clinical Neuropsychology, 16, 35–42.

Code, C. (1997) Can the right hemisphere speak? *Brain & Language, 57*, 38-39.

Darley, F.L., Aronson, A.E. & Brown, J.R. (1975). *Motor speech disorders*. Philadelphia: WB Saunders.

Dierckx, E., Ponjaert-Kristoffersen, I., Verté, D. & De Raedt, R. (2004). Use of neuropsychological test procedures to distinguish dementia from depression in the elderly. *Acta Neuropsychologica, 2(2)*, 130-141.

Esmonde ,T., Giles, E., Xuereb, J. & Hodges, J. (1996). Progressive supranuclear palsy presenting with dynamic aphasia. *Journal of Neurology, Neurosurgery and Psychiatry, 60(4)*, 403-10.

Garrard, P. & Hodges, J.R. (1999). Semantic dementia: implications for the neural basis of language and meaning. *Aphasiology, 13(8)*, 609-623.

Gates, G.A., Beiser, A., Rees, T.S., D'Agostino, R.B., Wolf, P.A. (2002), Central auditory dysfunction may precede the onset of clinical dementia in people with probable Alzheimer's disease. *Journal of the American Geriatric Society, 50*, 482–488.

Grossman, M., Rhee, J. & Moore, P. (2005). Sentence processing in frontotemporal dementia. *Cortex, 41*, 764-777.

Guiraud, P. (1956). *Psychiatrie clinique*. Paris: Le Francois.

Gustafson, L., Elfgren, C. & Passant, U. (2004). Fronto-temporal dementia: Differentiation from Alzheimer's disease. *Polish Journal of Geriatric Psychiatry, 1(4)*, 279-292.

Harciarek, M. & Jodzio, K. (2005). Neuropsychological differences between frontotemporal dementia and Alzheimer's disease: a review. *Neuropsychology Review, 15(3)*, 131-145.

Harciarek, M. & Kertesz, A. (2011). Primary progressive aphasias and their contribution to contemporary knowledge about the brain-language relationship. *Neuropsychology Review, Aug 2*. [Epub ahead of print].

Henry, J.D., Crawford, J.R. & Phillips, L.H. (2004). Verbal fluency performance in dementia of the Alzheimer's type: a metanalysis. *Neuropsychologia, 42(9)*, 1212-1222.

Hodges, J.R., Patterson, K., Oxbury, S. & Funnell, E. (1992). Semantic dementia: progressive fluent aphasia with temporal lobe atrophy. *Brain, 115*, 1783-1806.

Hughes, C. P., Berg, L., Danziger, W. L., Coben, L. A., & Martin, R. L. (1982). A new clinical scale for the staging of dementia. *The British Journal of Psychiatry*, *140*(6), 566.

Ioannides, P., Karacostas, D., Hatzipantazi, M. & Ioannis, M. (2005). Primary progressive aphasia as the initial manifestation of corticobasal degeneration: a "three in one" syndrome? *Functional Neurology, 20(3)*, 135-7.

Jackendoff, R. (2000). Fodorian modularity and representational modularity. In: Y. Grodzinsky, L.P. Shapiro & D. Swinney (eds.), *Language and the brain: representation and processing* (pp. 3-30). London: Academic Press.

Kaplan, E., Goodglass, H. & Weintraub, S. (1983). *The Boston Naming Test*. Philadelphia: Lea and Febiger.

Kempler, D. (1995). Language changes in dementia of the Alzheimer type. In: R. Lubinski (ed.), *Dementia and communication* (pp. 98-114). Los Angeles: Singular Publishing.

Kertesz, A. (1998a). Pick's disease and Pick's complex: introductory nosology. In: A. Kertesz & D.G. Munoz (eds.), *Pick's disease and Pick's complex* (pp. 1-11). New York: Wiley-Liss.

Kertesz, A. (1998b). Arnold Pick: a historical introduction. In: A. Kertesz & D.G. Munoz (eds.), *Pick's disease and Pick's complex* (pp. 13-21). New York: Wiley-Liss.

Kertesz, A. (2003). Pick complex: An integrative approach to frontotemporal dementia: primary progressive aphasia, corticobasal degeneration, and progressive supranuclear palsy. *Neurologist, 9*, 311-317.

Kertesz, A. (2005). Frontotemporal dementia: one disease, or many? Probably one, possibly two. *Alzheimer's Disease and Associated Disorders, 19 Suppl 1*, S19-24.

Kertesz, A., Davidson, W. & Fox, H. (1997). Frontal Behavioral Inventory: diagnostic criteria for frontal lobe dementia. *Canadian Journal of Neurological Sciences, 24,* 29-36.

Kertesz, A., Morlog, D., Light, M., Blair, M., Davidson, W., Jesso, S., & Brashear, R. (2008). Galantamine in frontotemporal dementia and primary progressive aphasia. *Dementia & Geriatric Cognitive Disorders, 25*, 178-85.

Kertesz, A, Blair, M., McMonagle, P. & Munoz, D.G. (2007). The diagnosis and course of frontotemporal dementia. *Alzheimer Disease & Associated Disorders, 21*, 155-63.

Kertesz, A., Jesso, S., Harciarek, M., Blair, M. & McMonagle, P. (2010). What is semantic dementia? A cohort study of diagnostic features and clinical boundaries. *Archives of Neurology, 67(4)*, 483-489.

Kertesz, A, Nadkarni, N., Davidson, W. & Thomas, A.W. (2000). The Frontal Behavioral Inventory in the differential diagnosis of frontotemporal dementia. *Journal of the International Neuropsychological Society, 6,* 460–68.

Knibb, J.A. & Hodges, J.R. (2005). Semantic dementia and primary progressive aphasia: a problem of categorization? *Alzheimer Disease and Associated Disorders, 19 Suppl 1*, S7-14.

Knopman, D.S., Mastri, A.R., Frey, W.H., Sung, J.H. & Rustan, T. (1990). Dementia lacking distinctive histologic features: a common non-Alzheimer degenerative dementia. *Neurology, 40(2)*, 251-256.

Lebrun, Y. (2003). A.R. Luria and non-volitional verbal behaviour. *Acta Neuropsychologica, 1(1)*, 22-33.

Lebrun, Y., Devreux, F. & Rousseau, J.J. (1986). Language and speech in a patient with a clinical diagnosis of progressive supranuclear palsy. *Brain & Language*, *27*, 247-256.

Leszek, J., Malyszczak, K., Janicka, B., Kiejna, A. & Wiak, A. (2003). Total tau in cerebrospinal fluid differentiates Alzheimer's disease from vascular dementia. *Medical Science Monitor, 9(11)*, CR484-488.

Litvan, I., Agid, Y., Calne, D. et al. (1996). Clinical research criteria for the diagnosis of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome): report of the NINDS-SPSP international workshop. *Neurology, 47(1)*, 1-9.

Lund and Manchester Groups (1994). Clinical and neuropathological criteria for frontotemporal demntia. *Journal of Neurology, Neurosurgery & Psychiatry, 57*, 416-418.

Luria, A.R. (1977). *Neuropsychologial studies in aphasia*. Neurolinguistics 6. Amsterdam: Swets & Zeitlinger B.V.

Luzzatti, C. & Poeck, K. (1991). An early description of slowly progressive aphasia. *Archives of Neurology, 48*, 228–229.

MacQueen, B.D. (2003). How a sentence unfolds. *Acta Neuropsychologica, 1(4)*, 382-406.

Mendez, M.F. & Cummings, J.L. (2003). *Dementia: a clinical approach*. 3rd ed. Philadelphia: Butterworth-Heinemann (Elsevier).

Mesulam, M.M. (1982). Slowly progressive aphasia without generalised dementia. *Annals of Neurology, 11*, 592–598.

Mesulam, M.M. (2001). Primary progressive aphasia. *Annals of Neurology, 49*, 425–432.

Miller, B.L., Boone, K., Mishkin, F., Schwartz, J.R., Koras, N. & Kushii, J. (1998). Clinical and neuropsychological features of frontotemporal dementia. In: A. Kertesz & D.G. Munoz D.G. (eds.) *Pick's disease and Pick's complex* (pp. 23-32). New York: Wiley-Liss.

Neary, D., Snowden, J.S., Northen, B. & Goulding, P.J. (1998). Dementia of frontal lobe type. *Journal of Neurology, Neurosurgery & Psychiatry, 51(3)*, 353-61.

Pachalska, M., Bidzan, L., Łukowicz, M., Bidzan, M., Markiewicz, K., Jastrzebowska, G. & Talar, J. (2011). Differential diagnosis of behavioral variant of fronto-temporal dementia (bvFTD). *Medical Science Monitor, 17(6)*, CR311-321.

Pąchalska, M. & MacQueen, B.D. (1998). *Kwestionariusz Zachowań Osoby z Zespołem Czołowym (BNT).* Kraków: Fundacja na Rzecz Osób z Dysfunkcjami Mózgu.

Pachalska, M., MacQueen, B. D. (2002). The collapse of the US-THEM structure in aphasia: a neuropsychological and neurolinguistic perspective. In A. Duszak (Ed.), Us & others. Social identities across languages, discourses, and cultures (pp. 481-503) . Amsterdam: John Benjamins.

Pąchalska, M., Kurzbauer, H., Grochmal-Bach, B., MacQueen, B.D., Urbanik, A. & Herman-Sucharska, I. (2003). Nietypowe zaburzenia języka i mowy u pacjentki z klinicznym rozpoznaniem zespołu Steele'a Richardsona-Olszewskiego. In: J. Leszek (red.), *Choroby otępienne: Teoria i praktyka* (pp. 285-304). Wrocław: Wyd. Continuo.

Pąchalska, M. (2008). *Rehabilitacja neuropsychologiczna: procesy poznawcze i emocjonalne*. Lublin: Wydawnictwo Naukowe UMCS.

Pasquier, F. & Delacourte, A. (1998). Non-Alzheimer degenerative dementias. *Current Opinion in Neurology, 11*, 417-427.

Ross, G.W., Cummings, J.L. & Benson, D.F. (1990). Speech and language alterations in dementia syndromes: characteristics and treatment. *Aphasiology, 4*, 339-352.

Selnes, O.A. & Harciarek, M. (2005). Primary progressive aphasia without dementia. In: K. Brown (ed.), *The encyclopedia of language and linguistics*, 2nd edition (pp. 88-91). Amsterdam: Elsevier Science.

Snowden, J.S. & Neary, D. (1993). Progressive language dysfunction and lobar atrophy. *Dementia, 4(3)*, 226-231.

Stern, Y. (2002). What is cognitive reserve? Theory and research application of the reserve concept. *Journal of the International Neuropsychological Society, 8*, 448-460.

Stern, Y., Zarahn, E., Hilton, H.J., Flynn, J., DeLaPaz, R. & Rakitin, B. (2003). Exploring the neural basis of cognitive reserve. *Journal of Clinical and Experimental Neuropsychology, 25(5)*, 691-701.

Varma, A.R., Adams, W., Lloyd, J.J., Carson, K.J., Snowden, J.S., Testa, H.J. et al. (2002). Diagnostic pattern of regional atrophy on MRI and regional cerebral blood flow change on SPECT in young onset patients with Alzheimer's disease, frontotemporal dementia and vascular dementia. *Acta Neurologica Scandinavica, 105*, 261-269.

Address for correspondence:
Prof. Maria Pachalska, M.D., Ph.D.
Chair of Neuropsychology
Andrzej Frycz-Modrzewski Kraków University
ul. Herlinga-Grudzinskiego 1, 30-705 Krakow, Poland
e-mail: neuropsychologia23@o2.pl