We report five patients with a stereotyped clinical syndrome characterised by fluent dysphasia with severe anomia, reduced vocabulary and prominent impairment of single-word comprehension, progressing to a stage of virtually complete dissolution of the semantic components of language. A marked reduction in the ability to generate exemplars from restricted semantic categories (e.g. animals, vehicles etc.) was a consistent and early feature. Tests of semantic memory demonstrated a radically impoverished knowledge about a range of living and man-made items. In contrast, phonology and grammar of spoken language were largely preserved, as was comprehension of complex syntactic commands. Reading showed a pattern of surface dyslexia. Autobiographical and day-to-day (episodic) memory were relatively retained. Non-verbal memory, perceptual and visuospatial abilities were also strikingly preserved. In some cases, behavioural and personality changes may supervene; one patient developed features of the Kluver-Bucy Syndrome. Radiological investigations have shown marked focal temporal atrophy in all five patients, and functional imaging by SPECT and PET (one case) have implicated the dominant temporal lobe in all five. In the older literature, such cases would have been subsumed under the rubric of Pick's disease. Others have been included in series with progressive aphasia. We propose the term semantic dementia, first coined by Snowden et al. (1989) to designate this clinical syndrome.

A patient, JL, with the syndrome of semantic dementia was assessed longitudinally over a two-year period. The data presented here address the controversy concerning the hierarchical organisation of semantic memory. On a range of category fluency tests, when first tested JL was just within the normal range on the broadest categories of animals and household items, but was virtually unable to produce any instances of specific categories such as breeds of dog or musical instruments. Longitudinal fluency data for the animal category demonstrate that while JL continued to produce the most prototypic responses (cat, dog, horse), other animal labels dropped out early from his vocabulary. On the picture sorting tests from our semantic memory test battery, JL's discrimination between living things and manmade objects was preserved for a substantial time in conjunction with a marked decline in his sorting ability for more specific categories, particularly features or attributes (e.g., size, foreign-ness or
ferocity of animals). An analysis of naming responses to the 260 Snodgrass and Vanderwart pictures on four occasions suggests a progressive loss of the features of semantic representations that enable discrimination between specific category instances. There was a progressive decline in circumlocutory and category co-ordinate responses with a rise in broad superordinate and cross-category errors. The latter are of particular theoretical interest; on session I, all cross-category errors respected the living/manmade distinction, but by session IV almost half of such errors failed to respect this distinction. The emergence of category prototypes was another notable feature, particularly in the living domain: at one stage, land (or four-legged) animals were all named either cat, dog or horse. By contrast, within the manmade domain, items were frequently described in terms of their broad use or function, until eventually no defining features were produced. These findings are discussed in the context of competing theories of semantic organisation.

Authors: Moss, H.E., Tyler, L.K., Hodges, J.R., Patterson, K.
Title: Exploring the loss of semantic memory in semantic dementia: Evidence from a primed monitoring study
Reference: Neuropsychology, 9, 16-26
Year: 1995
Key Words: neuropsychology, semantic memory, priming
Abstract:

We used an on-line primed monitoring study to explore the nature of semantic memory loss in a patient (PP) with semantic dementia, who showed a profound semantic impairment on a range of off-line tasks. Priming for pairs of words taken from a common category (e.g. cat-dog, spade-rake, ruby-emerald) was contrasted with that for word pairs from different categories which were related functionally (e.g. shampoo-hair, broom-floor, theatre-play). Control subjects showed robust priming for both types of semantic relation. PP, in contrast, showed a normal priming effect for the functionally related conditions but no priming for category co-ordinates. This suggests that PP's semantic memory loss cannot be explained as either one of loss of stored representations or a problem with deliberate controlled access to that information, but has elements of both for different kinds of semantic information.

Authors: Hodges, J.R., Patterson, K.
Title: Non-fluent progressive aphasia and semantic dementia: A comparative neuropsychological study.
Reference: Journal of the International Neuropsychological Society, 2, 511-524
Year: 1996
Key Words: neuropsychology; aphasia; language; semantic memory
Abstract:

Two patients with non-fluent progressive aphasia, who have been studied longitudinally, are contrasted with a group of five patients with fluent progressive aphasia or semantic dementia. The most prominent feature of non-fluent syndrome is the severe distortion of speech output with phonological errors and agrammatic sentence structure. This contrasts with fluent, articulated and syntactically correct, but empty anomic speech found in semantic dementia. Performance on tests of comprehension separates the patient groups; the non-fluent patients show normal single-word comprehension but marked impairment on tests of syntactic comprehension, while those with semantic dementia demonstrate the opposite pattern. Category fluency is severely defective in semantic dementia, but initial letter-based fluency is more impaired in the non-fluent syndrome. Performance on non-verbally mediated tests of semantic knowledge is impaired in semantic dementia only. The two forms of progressive aphasia
have in common the sparing of perceptual and visuo-spatial skills, non-verbal problem solving abilities and day-to-day (episodic) memory. Neuroradiological investigations have shown marked selective and striking infero-lateral left temporal lobe atrophy in all five patients with semantic dementia: the changes in non-fluent progressive aphasia appear to be less focal and involve left perisylvan structure more diffusely. These two forms of progressive aphasia are, we argue, distinct in their manifestations.

Authors : Graham, K.S., Patterson, K., Hodges, J.R.
Title : Semantic dementia and pure anomia: two varieties of progressive fluent aphasia.
Year : 1998
Key Words : Progressive aphasia, semantic memory, semantic dementia, anomia, neuropsychology
Abstract :

We compared the performance of one patient with typical semantic dementia (JL) with that of another patient (FM) who seemed to represent a different variety of progressive fluent aphasia. A series of neuropsychological and experimental investigations suggested that FM's anomia was not due to a loss of semantic knowledge or an impairment at the phonological level. These studies found the following: (1) Over time (3 years of longitudinal testing) FM had only a mild and stable impairment on tests of semantic memory, unlike other patients with semantic dementia, who often show rapid decline on such tests. (2) While FM performed at ceiling on tests of immediate single word repetition, she often lost the phonological form of words when a delay was introduced between her hearing the word and attempting to produce it. By contrast, her ability to perform word-picture matching tests was not affected by a timed delay, suggesting that she still had access to semantic information about the spoken word. (3) By using two gating experiments in which FM heard and had to identify incrementing fragments of concrete words, she showed near normal performance and minimal benefit from the provision of a semantic clue in the form of a picture of the spoken word. These results suggest that FM's deficits were due to a loss of communication between semantic memory and the phonological output lexicon: a case of progressive pure anomia rather than semantic dementia.

Authors : GRAHAM, K.S., Becker, J.T., HODGES, J.R.
Title : On the relationship between knowledge and memory for pictures: Evidence from the study of patients with semantic dementia and Alzheimer's disease.
Year : 1997
Abstract :

Current views of long-term memory presume that both the hippocampal complex and the neocortex play interactive, but separate, roles in the storage of memories. While the neocortex is considered the eventual and permanent store for our memories, the encoding of recently experienced events is thought to be initially dependent upon the hippocampus and closely related structures. Neuropsychological studies have demonstrated that damage to the medial temporal lobe results in a retrograde amnesia extending back in time, with better preservation of older memories. The converse pattern has been shown in patients with semantic dementia, who have focal atrophy of the inferolateral temporal neocortex, but relative sparing of the hippocampal complex (Graham & Hodges, 1997). Here we demonstrate that such patients can show relatively preserved new learning on a forced-choice recognition memory test (based on real and chimeric animals), while patients in the early amnesic phase of Alzheimer's disease show severely impaired learning on the same test. This result provides
support for the view that new learning is primarily dependent upon the hippocampus and related structures.

Authors: GRAHAM, K.S., Lambon Ralph, M.A., HODGES, J.R.
Title: Determining the impact of autobiographical experience on “Meaning”: New insights from investigating sports-related vocabulary and knowledge in two cases with semantic dementia.
Reference: Cognitive Neuropsychology, 14, 801-837.
Year: 1997
Key Words: autobiographical memory, semantic memory, progressive aphasia

Abstract:

Snowden, Griffiths, and Neary (1994, 1995) have proposed that autobiographical experience helps to maintain the integrity of semantic memory in patients with semantic dementia. We investigated this hypothesis by testing knowledge related to golf and bowls in two case studies. If Snowden and colleagues’ hypothesis is correct, our two patients should have better semantic knowledge for the sport that they regularly experience, compared with knowledge of other sports. In keeping with Snowden et al.’s hypothesis, we found that autobiographical experience influenced the ability of the patients to match up a surname with a first name: The names of personally and currently relevant golf/bowls partners were more likely to be matched correctly than such personally relevant names from the past, or the names of famous sports celebrities. Unlike Snowden et al., however, we found that knowledge of people, in all categories, was severely impoverished and that any semantic information was produced as part of an autobiographical memory. Likewise, detailed study of each patient’s understanding of their favourite sport revealed no significant effect of autobiographical experience on true semantic knowledge. We propose that the ability of semantic dementia patients to encode, albeit temporarily, recent autobiographical memories via a spared hippocampal complex supports the production of highly autobiographically constrained semantic-like facts and, to a lesser extent, frequently encountered names. There is, however, no direct effect of autobiographical experience on previously established semantic memory, i.e. knowledge of golf, bowls, and people, presumably stored within the temporal neocortex. These results are discussed with respect to current anatomically based computational models of long-term memory.

Authors: HODGES, J.R., GRAHAM, K.S.
Title: A reversal of the temporal gradient for famous person knowledge in semantic dementia: implications for the neural organisation of long-term memory.
Reference: Neuropsychologia, 36, 803-825
Year: 1998

Abstract:

On tests of autobiographical memory, patients with semantic dementia demonstrate significantly better retrieval of episodic events from the recent past compared with the distant past. This reversal of the Ribot effect has been attributed to the relative sparing of the hippocampal complex in the disorder. Current computational models of long-term memory predict a similar time-dependent pattern of impairment on tests of remote semantic memory. Five patients with semantic dementia were tested on recognition (familiarity) and identification (knowledge) of famous names selected from four different time-periods: 1950’s, 1980’s, 1990-1993 (early 1990’s) and 1994-1996 (current). As expected, it was found that while DM (who had focal left temporal lobe atrophy) showed no significant impairment on recognition of famous names, he was significantly better at producing information about people who were currently famous compared to people famous in the other three time-periods. The other four patients (who had bilateral temporal lobe damage) showed better recognition of famous names from the
current time-period (and to a lesser extent the 1950's), yet were profoundly impaired on the
identification component, producing very little information across all four time periods. The results are
discussed with respect to current views of the neural organisation of person-specific and general
semantic memory.

Authors: LAMBON RALPH, M.A., GRAHAM, K.S., Ellis, A.W., HODGES, J.R.
Title: Naming in semantic dementia - what matters?
Year: 1998
Key Words: Progressive aphasia, naming, speech production

Abstract:

One of the major symptoms of semantic dementia (or progressive fluent aphasia) is profound word-
finding difficulties. We present here a cross-sectional study of the factors affecting picture naming in
semantic dementia based on data obtained from eight patients, together with a longitudinal analysis of
naming in another patient. Various properties and attributes of the objects were entered into a series of
regression analyses in order to predict which items the patients could or could not name. The analyses
showed that object familiarity, word frequency and age-of-acquisition predicted naming success for the
group and, in most cases, for each individual patient, irrespective of lesion site or overall naming
success. We propose that the pattern of naming in semantic dementia is best described in terms of
reduced semantic activation within a cascading/interactive speech production system. We suggest that
object familiarity, and possibly word frequency, reflect the inherent robustness of individual semantic
representations to the decay process in terms of both quantity and quality of experience. Age-of-
acquisition and word frequency (at a phonological-lexical level) predicts naming success, because
frequent, early-acquired words are relatively easy to activate even with reduced semantic 'input'.

Authors: Knott, R., PATTERSON, K., HODGES, J.R.
Title: Lexical and semantic binding effects in short-term memory: Evidence from semantic dementia.
Reference: Cognitive Neuropsychology, 14, 1165-1216.
Year: 1997
Key Words: Memory: short-term memory, semantic memory; Language: lexical processes, speech
production; Neuropsychology: progressive aphasia, semantic memory

Abstract:

Two case studies are presented of the short-term memory performance of patients with semantic
dementia. The first case showed a pervasive pattern of semantic effects in his auditory verbal short-
term memory performance. In particular, a marked superiority in serial recall for sequences of
Known words over Unknown words - words which he no longer comprehended. His performance
in serial recall tasks was characterised by abundant phonemic errors which occurred with a frequency
shown to be related to semantic factors. These errors were often migrations of phonemes from one
word in the list to another, suggesting that impaired semantic processing reduces the Glue or
Binding that helps to maintain a structured sequence of phonemes in short-term memory. The
second patient also showed some semantic effects in serial recall, and a significant but less marked
pattern of phonological errors. The differing performance of the two patients is interpreted within an
interactive activation model of word production (Saffran and Martin, 1990; Martin, Saffran & Dell,
1996).

Authors: HODGES, J.R., Garrard, P., PATTERSON, K.
Title: Semantic Dementia.
In tracing the origins of the term semantic dementia, it is necessary to draw on three initially separate bodies of neurological and neuropsychological literature: those relating to Pick’s disease, progressive aphasia, and semantic memory impairment. The authors cover clinical features, prognosis, radiological findings and experimental aspects of semantic dementia.

Authors: GRAHAM, K.S., Pratt, K.H., HODGES, J.R.

Title: A reverse temporal gradient for public events in a single case of semantic dementia

Abstract:

A patient with semantic dementia (DM) was tested on a Public Events Test comprising famous events from three time periods (1989-1991, 1992-1994, 1nd 1995-1997). DM was compared on both recognition and identification of these events compared to two matched control subjects. Furthermore, he showed evidence of a reverse temporal gradient on both components of the test, recognizing more items and producing more detailed information about events from 1995 to 1997 compared to events from the other two time periods. The results from this experiment are congruent with data from other published studies in which patients with semantic dementia show reverse temporal gradients on tests of autobiographical and semantic knowledge.

Authors: HODGES, J., PATTERSON, K., WARD, R., Garrard, P., BAK, T., Perry, R., and Gregory, C.

Title: The differentiation of semantic dementia and frontal lobe dementia (temporal and frontal variants of frontotemporal dementia) from early Alzheimer’s Disease: a comparative neuropsychological study.

Abstract:

The authors compared age-matched groups of patients with the frontal and temporal lobe variants of frontotemporal dementia (FTD; dementia of frontal type [DFT] and semantic dementia), early Alzheimer’s disease (AD), and normal controls (n = 9 per group) on a comprehensive neuropsychological battery. A distinct profile emerged for each group: Those with AD showed a severe deficit in episodic memory with more subtle, but significant, impairments in semantic memory and visuospatial skills; patients with semantic dementia showed the previously documented picture of isolated, but profound, semantic memory breakdown with anomia and surface dyslexia but were indistinguishable from the AD group on a test of recall; and the DFT group were the least impaired and showed mild deficits in episodic memory and verbal fluency but normal semantic memory. The frontal and temporal presentations of FTD are clearly separable from each other and early AD.

Authors: GRAHAM, K.S., PATTERSON, K., HODGES, J.

Title: Episodic memory: new insights from the study of semantic dementia
The study of patients with semantic dementia, the temporal variant of frontotemporal dementia, has provided new insights into the interaction between episodic and semantic memory, and the different roles played by various structures in the temporal lobe. Recent findings indicate that the syndrome of semantic dementia can inform us about the organisation of long-term memory and the relationship between semantic memory and other cognitive systems.

Authors: HODGES, J.R
Title: Memory in the dementias: with special reference to Alzheimer's disease and semantic dementia.

This chapter provides a comprehensive review of episodic and semantic memory in a range of dementing illnesses, but highlights advances over the past decade in the areas of Alzheimer's disease and semantic dementia.

Authors: HODGES, J.R
Title: Pick's Disease: It's relationship to progressive aphasia, semantic dementia and frontotemporal dementia.

The aims of this chapter is to review the evolution of the terms used to describe this spectrum of disorders, to highlight recent advances and areas of continuing controversy.

Authors: GRAHAM, K.S., Murre, J.M.J., HODGES, J.R.
Title: Episodic memory in semantic dementia: A computational approach based on the TraceLink model.

In this chapter, we discuss how the neuropsychological characteristics of semantic dementia can inform us about the roles of the temporal neocortex and the hippocampal complex in the acquisition and storage of episodic and semantic memory. In particular, we review a computational model of amnesia, TraceLink (Murre, 1994, 1996, 1997), and determine whether predictions from this model are applicable to the syndrome of semantic dementia.
Aggleton and Brown propose that the hippocampal-0anterior thalamic and perirhinal-medial dorsal thalamic systems play independent roles in episodic memory, with the hippocampus supporting recollection-based memory and the perirhinal cortex recognition memory. In this commentary we discuss whether there is experimental support for Aggleton and Brown's model from studies of long-term memory in semantic dementia.
Volumetric analysis of MRI scans suggests the perirhinal cortex may be preserved in semantic dementia. Recent findings of preserved non-verbal new episodic learning in the disorder imply that the perirhinal cortex may be implicated in recognition memory, and not the processing of semantic knowledge as has been claimed.

Authors: PATTERSON, K., HODGES, J.R.
Title: *Semantic dementia: One window on the structure and organisation of semantic memory.*

Abstract:

This chapter concerns one of the most central of human cognitive abilities, semantic memory, viewed from the perspective of the neuropsychological condition in which this centre falls apart. The chapter begins with summaries of the cognitive, neuroanatomical and pathological features of semantic dementia, and then reviews recent research addressed to two questions: (i) Is semantic memory one or several systems? Differences associated with the contributions of left- and right-temporal lobes to conceptual knowledge; (ii) Is semantic memory one or several systems? Differences associated with words vs objects as input, and with speech vs action as output.

Authors: Lambon Ralph, M.A., Graham, K.S., Patterson, K., Hodges, J.R.
Title: *Is a picture worth a thousand words? Evidence from concept definitions by patients with semantic dementia.*

Abstract:

Nine patients with semantic dementia (the temporal lobe variant of frontotemporal dementia) were asked to define concrete concepts either from presentation of a picture of the object or its spoken name. As expected, the patients with the most severe semantic impairment produced the least detailed definitions, and the quality of the definitions overall was significantly related to concept familiarity. Further analyses of the definitions were designed to assess two key theoretical aspects of semantic organization. (i) Do objects and their corresponding names activate conceptual information in two neuroanatomically separable (modality-specific) semantic systems? If so, then - apart from any expected commonality in performance attributable to factors such as concept familiarity - one would not predict striking item-specific similarities in a patient's picture- and word-elicited definitions. (ii) Do sensory/perceptual features and more associative/functional attributes of conceptual knowledge form two neuroanatomically separable sub-systems? If so, then one would predict significant dissociations in the prominence of these two types of information in the patients' definitions. The results lead us to favour a model of the semantic system that is divided by attribute type but not by modality.

Authors: Mummery, C.J., PATTERSON, K., Price, C.J., Ashburner, J., Frackowiak, R.S.J., HODGES, J.R.
Title: *A voxel based morphometry study of semantic dementia: The relationship between temporal lobe atrophy and semantic dementia*
The cortical anatomy of six patients with semantic dementia (the temporal lobe variant of frontotemporal dementia) was contrasted with that of a group of age-matched normals using voxel-based morphometry, a technique that identifies changes in grey matter volume on a voxel-by-voxel basis. Among the circumscribed regions of neuronal loss, the left temporal pole (BA 38) was the most significantly and consistently affected region. Cortical atrophy in the left hemisphere also involved the infero-lateral temporal lobe (BA 20/21) and fusiform gyrus. In addition, the right temporal pole (BA 38), the ventromedial frontal cortex (BA 11/32) bilaterally and the amygdaloid complex were affected, but no significant atrophy was measured in the hippocampus, entorhinal or caudal perirhinal cortex. The degree of semantic memory impairment across the six cases correlated significantly with the extent of atrophy of the left anterior temporal lobe but not with atrophy in the adjacent ventromedial frontal cortex. These results confirm the view that the anterior temporal lobe is critically involved in semantic processing, and dissociate its function from that of the adjacent frontal region.

Authors: Garrard, P., Hodges, J.R.
Title: Semantic Dementia: Clinical, radiological and pathological perspectives
Reference: Journal of Neurology, 247 (6), 409-422
Year: 2000
Abstract:

Semantic dementia is a recently described clinical syndrome, characterised by an acquired progressive inability to name or comprehend common concepts, with little or no distortion of the phonological and syntactic aspects of language, and relative sparing of other aspects of cognition, such as episodic memory, non-verbal problem solving, and perceptual and visuo-spatial skills. The cognitive locus of this syndrome appears to lie in the permanent store of long-term memory representing general word knowledge - semantic memory. The anatomical distribution of atrophy is less well defined, and the contribution of various imaging modalities is discussed in the context of a body of 45 published and unpublished cases. We conclude that involvement of the left inferolateral temporal cortex is the critical area in the genesis of SD. SD probably always represents a non-Alzheimer neurodegenerative process; a variety of pathological lesions may be present, and possible aetiologies, together with debates about their correct classification, are discussed.

Authors: HODGES, J.R.
Title: Semantic Dementia: a selective disorder of semantic memory?
Reference: DÕEsposito (Ed) - Cognitive Neuroscience: a neurological perspective
Year: 1999
Abstract:

The chapter reviews work by our group on the topic of semantic dementia over the past decade with an emphasis on the insights that this work has given us into the organisation of semantic memory

Authors: BOZEAT, S., LAMBON RALPH, M.A., PATTERSON, K., Garrard, P., HODGES, J.R.
Title: Non-verbal semantic impairment in semantic dementia
Reference: Neuropsychologia 2000, 38 (9), 1207-1215
The clinical presentation of patients with semantic dementia is dominated by anomia and poor verbal comprehension. Few studies have investigated semantic impairment using entirely non-verbal assessments and the few exceptions have been based on results from single cases (Breedin et al., 1994; Graham et al., 1997; Lambo Ralph & Howard, in press). This study employed sound recognition and semantic association tasks to investigate the nature of the verbal and non-verbal comprehension deficit in ten patients with semantic dementia. The patients were impaired on both verbal and non-verbal assessments, and their accuracy on these tasks was directly related to their scores on a range of other tests requiring access to semantic memory. Further analyses revealed that performance was graded by concept and sound familiarity and, in addition, identified significant item consistency across the different conditions of the tasks. These results support the notion that the patients’ deficits across all modalities were due to degradation within a single, central network of conceptual knowledge. There were also reliable differences between conditions, which we propose arise directly from the nature of the mapping between input modality and semantic memory. Words and sounds have an arbitrary relationship with meaning while pictures benefit from a degree of systematicity with conceptual knowledge about the object.

Authors: SIMONS, J.S., GRAHAM, K.S., PATTERSON, K., HODGES, J.R.
Title: Multiple inputs to episodic memory: Evidence from semantic dementia (Abstract).
Reference: Hippocampus

We compared recognition memory for pictures of 'known' and 'unknown' objects and faces in three patients with semantic dementia. All three patients showed preserved recognition memory for the pictures if they were identical at study and test, irrespective of the status of their semantic knowledge about the target items. A direct correspondence between conceptual knowledge of, and recognition memory for, a familiar object or famous person was seen only when the item used at study was replaced with a perceptually different exemplar (e.g., a different photograph of the object/person) in the test phase of the episodic task. These results are inconsistent with Tulving's (1995) model of long-term memory. We propose instead that the data from semantic dementia support a model in which new episodic learning can draw upon perceptual information (e.g., vision, audition, etc.) and semantic knowledge.

Authors: HODGES, J.R., BOZEAT, S., PATTERSON, K., Spatt, J.
Title: The Role of Conceptual Knowledge in Object Use: evidence from semantic dementia.

It has been reported that patients with semantic dementia function well in everyday life and sometimes show striking preservation of the ability to use objects, even those specific objects for which the patient has degraded conceptual information. To explore this phenomenon in nine cases of semantic dementia, we designed a set of semantic tests regarding 20 everyday objects and compared performance on these with the patients' ability to demonstrate the correct use of the same items. We also administered a test of mechanical problem solving utilising novel tools (from Goldenberg and Hagmann, 1998), on which the patients had completely normal ability. All but the mildest patient showed significant deficits of
naming and on the visually-based semantic matching tasks. Object use was markedly impaired and, most importantly, correlated strongly with naming and semantic knowledge. In a small number of instances, there was appropriate use of an object for which the patient's knowledge on the semantic matching tasks was no better than chance; but this typically applied to objects with a rather obvious relationship between appearance and use, or was achieved by trial and error. The results suggest that object use is heavily dependent upon object-specific conceptual knowledge, supplemented to some degree by a combination of visual affordances and mechanical problem solving.

Authors: BIRD, H.E., LAMBON RALPH, M.A., PATTERSON, K., HODGES, J.R.
Title: The Rise and Fall of Frequency and Imageability: Noun and Verb Production in Semantic Dementia
Reference: Brain and Language, 73, 17-49
Year: 2000
Key Words: semantic dementia, progressive aphasia, nouns, verbs, frequency, imageability, narrative, spontaneous speech, Cookie Theft, picture description
Abstract:

This study examines the impact of progressive degeneration of conceptual knowledge on the content words used in connected speech elicited using the Cookie Theft picture description (Goodglass & Kaplan 1983). We began with an analysis of control subjects' descriptions with regard to word types and their frequency and imageability. Because the impairment of conceptual knowledge in semantic dementia is graded by concept familiarity, we created a model of a standardised normal Cookie Theft description that was then progressively degraded by the systematic removal of lower bands of word frequency. We drew two main predictions from this model: reduced availability of the lower bands of word frequency should result in (a) an apparent deficit for noun retrieval in relation to verb retrieval, and (b) an apparent reverse imageability effect. Results from a longitudinal study, in which three patients with semantic dementia each described the Cookie Theft picture on three occasions during the progression of their disease, confirmed these predictions. An additional cross-sectional analysis, adding narratives from a larger number of cases, demonstrated that the decline in ability to produce suitable words for the picture description is closely related to the extent of semantic impairment as measured in tests of word comprehension and production. Both verbs and nouns are affected by the degradation of semantic memory; the fact that the impairment to noun production is manifested earlier and more catastrophically may be attributed to the relatively lower frequency of these terms.

Authors: SIMONS, J.S., GRAHAM, K.S., Galton, C.J., PATTERSON, K., HODGES, J.R.
Title: Semantic knowledge and episodic memory for faces in semantic dementia
Reference: Neuropsychology, 15 (1), 101-114
Year: 2001
Key Words: Episodic memory; semantic memory; recognition memory; frontotemporal dementia; temporal lobe; perirhinal cortex
Abstract:

Although semantic dementia has been characterised as a selective disorder of semantic knowledge with preserved episodic memory, previous studies have documented poor performance on Warrington's (1984) Recognition Memory Test for Faces. We demonstrated here, however, that so long as atrophy affected predominantly the left temporal lobe, patients with semantic dementia showed preserved face recognition memory. Patients with structural damage to the right temporal lobe were typically impaired, and analyses indicated that the status of the hippocampus and parahippocampal gyrus (including the perirhinal cortex) on the right was critical. Two single-case studies of patients with
predominantly left temporal lobe pathology confirmed that their recognition memory for photographs of famous faces was intact, even if their semantic knowledge about the celebrities depicted was severely degraded. An effect of semantic knowledge on recognition memory became apparent only when perceptually different photographs of the famous people were used in the study and test phases. These results support a view in which new episodic learning is typically supported by information from both perceptual and semantic systems.

Authors: BOZEAT, S., Lambon Ralph, M.A., PATTERSON, K.P., HODGES, J.R.
Title: The Influence of Personal Familiarity and Context on Object Use in Semantic Dementia
Reference: Neurocase, 8, 127-134
Year: 2002
Abstract:

Several previous studies of semantic dementia, involving formal assessment of the ability to demonstrate the use of individual objects, have revealed a significant deficit that corresponded directly to the patients’ remaining semantic knowledge for the same items. Until late in the course of disease, however, patients with semantic dementia are reported to demonstrate completely normal use of some objects that are relevant to their everyday living or hobbies. The aim of this study was to explore the apparent difference between formal assessment of object use in the clinic and informal observations of patients in their own homes using their own objects. Specifically, we examined the influence of two factors: personal familiarity with object exemplars and the contexts in which they are typically used. Two patients with severe semantic impairment were asked to demonstrate the use of a set of their own objects that they were still using at home on a regular basis. Performance on these items was compared with two additional sets: perceptually-similar and perceptually-different exemplars of the same objects. All three sets were tested both in the patients’ own homes and in a neutral environment. Both patients demonstrated better use of their own objects than of the perceptually different exemplars. An advantage for ‘own’ relative to perceptually similar exemplars characterised only one of the two patients. Neither patient showed a significant benefit of familiar home context on object use. This study confirms a discrepancy between impaired performance on objects selected for clinical assessment and relative preservation for a limited set of objects that are used for daily living. The results suggest that repeated experience with personally familiar objects helps to maintain appropriate responses to them in the face of severe degradation of conceptual knowledge.

Authors: Murre, J.M.J., GRAHAM, K.S., HODGES, J.R.
Title: Semantic dementia: relevance to connectionist models of long-term memory
Reference: Brain, 124, 647-675
Year: 2001
Key Words: connectionist models; hippocampus; long-term memory; memory consolidation; retrograde amnesia; semantic dementia
Abstract:

Semantic dementia is a recently documented syndrome associated with non-Alzheimer degenerative pathology of the polar and inferolateral temporal neocortex, with relative sparing (at least in the early stages) of the hippocampal complex. Patients typically show a progressive deterioration in their semantic knowledge about people, objects, facts and the meanings of words. Yet, at least clinically, they seem to possess relatively preserved day-to-day (episodic) memory. Neuropsychological investigations of semantic dementia provide, therefore, a unique opportunity to investigate the organization of human long-term memory and, more specifically, to determine the relationship between semantic memory and other cognitive systems, such as episodic memory. In this review, we summarize
recent empirical findings from patients with semantic dementia and discuss whether the neuropsychological phenomena of the disease are consistent with current cognitive and computational models of human long-term memory and amnesia. Six specific issues are addressed: (i) the relative preservation of category-level (superordinate) compared with fine-graded (subordinate) semantic knowledge as the disease progresses; (ii) the better recall of recent autobiographical and semantic memories compared with those in the distant past; (iii) the preservation of new learning, as measured by recognition memory, early in the disease; (iv) the interaction between autobiographical experience and semantic knowledge in the current, but not the distant, time-period; (v) increased long-term forgetting of newly learned material; and (vi) impaired implicit memory. It is concluded that recent findings from semantic dementia offer strong support for the view that memory consolidation in humans is dependent upon interactions between the hippocampal complex and neocortex. Furthermore, these investigations have provided computational modellers of human memory with a novel set of neuropsychological data to be simulated and tested.

Authors: Lambon Ralph, M.A., McClelland, J.L., Patterson, K., Hodges, J.R.
Reference: Higher Brain Function Research (Shitsugosho Kenkyu), 20 (2) 145-156
Year: 2000
Key Words: semantic dementia, semantic memory, speech production, connectionist PDP models, neuroanatomy
Abstract:

The relationship between conceptual knowledge and object naming was investigated in a study that combined a computational model of single word production with longitudinal and cross-sectional data of patients with semantic dementia (a selective decline of semantic memory resulting from progressive temporal-lobe atrophy). Although all patients with semantic dementia have both anomia and impaired comprehension, previous reports had indicated two different longitudinal profiles: (a) a parallel decline in accuracy of naming and comprehension with frequent semantic naming errors, suggesting a purely semantic basis for the anomia; (b) a dramatic progressive anomia without commensurate decline in comprehension, which might suggest a mainly post-semantic source of the anomia. Results, described here, for 16 patients with semantic dementia reflected these two profiles, but with the following important caveats: (1) despite a few relatively extreme versions of one or other profile, the full set of cases formed a continuum in the extent of anomia for a given degree of degraded comprehension; (2) the degree of disparity between these two abilities was associated with relative asymmetry in laterality of atrophy: a parallel decline in comprehension and naming characterized patients with greater right-than left-temporal atrophy, while disproportionate anomia occurred with a predominance of atrophy in the left temporal lobe. These patterns were successfully reproduced in an implemented computational model of naming. This model incorporated semantic representations that were distributed across simulated left-and right-temporal regions but the semantic units on the left were more strongly connected to left-lateralized phonological representations. Bilateral but asymmetric damage to semantic units replicated the longitudinal patient profiles of naming relative to comprehension. On the basis of both the neuropsychological and computational evidence, we propose that semantic impairment alone can account for the full range of word production deficits observed in semantic dementia.

Authors: Graham, K.S., Patterson, K., Pratt, K.H., Hodges, J.R.
Reference: Neuropsychological Rehabilitation, 11 (3/4), 429-454
Title: Can repeated exposure to 'forgotten' vocabulary help alleviate word-finding difficulties in semantic dementia? An illustrative case study.
The predominant, and most socially isolating, symptom typically seen in semantic dementia is anomia — word-finding difficulties — in conjunction with a deteriorating central semantic system. In this paper, we demonstrate that repeated rehearsal of the names of concepts paired with pictures of them and/or real items resulted in a dramatic improvement in the ability of a patient with semantic dementia (DM) to produce previously "difficult-to-retrieve" words on tests of word production. Although the substantial improvement shown by DM suggests that home rehearsal with pictorial and verbal stimuli could be a useful rehabilitative strategy for word-finding difficulties in semantic dementia, the experiment also revealed that constant exposure to items was necessary in order to prevent the observed decline in performance once DM's daily drill was stopped. The results are discussed with respect to the underlying neuroanatomical structures thought to be important for the acquisition and storage of long-term memory, and to techniques for facilitating word-finding in patients with aphasia.

Authors: HODGES, J.R., Miller, B.L.
Title: The neuropsychology of frontal variant frontotemporal dementia and semantic dementia. Introduction to the special topic papers: Part II.
Reference: Neurocase, 7(2), 113-121
Year: 2001
Abstract:

The second part of this review, which accompanies 10 special articles dedicated to frontotemporal dementia (FTD), describes some of the advances in understanding frontal variant (fvFTD) and semantic dementia, and the theoretical insights gained into normal cognitive processes from the study of these syndromes. Recent work has clarified the neuropsychiatric features that distinguish fvFTD from Alzheimer's and has begun to spread light on the underlying deficits in social judgement, theory of mind, processing of emotional stimuli and decision making. With regard to central aspects of cognitive processing, such as memory, patients with fvFTD may have a distinctive profile of performance on tests of explicit and implicit memory, and contrary to current views some patients may even have a severe amnesic syndrome. There is also growing evidence that despite relatively well preserved general language skills, patients with fvFTD have particular problems with verb processing. Although there is agreement regarding the key features of semantic dementia, much controversy has surrounded the issue of whether the cognitive findings favour an explanation in terms of progressive breakdown of a central amodal semantic store or differential involvement of verbal and non-verbal systems. Other topics discussed include the impact of semantic breakdown on phonological processes and episodic memory. We also review recent structural and functional neuroimaging findings in semantic dementia.

Authors: HODGES, J.R., GRAHAM, K.
Title: Episodic Memory: insights from semantic dementia.
Reference: Phil. Trans. R. Soc. Lond. B, 356, 1423-1434
Year: 2001
Abstract:

Semantic dementia, also known as the temporal lobe variant of frontotemporal dementia, results in a progressive yet relatively pure loss of semantic knowledge about words, objects and people, and is associated with asymmetric, focal atrophy of the antero-lateral temporal lobes. Semantic dementia provides a unique opportunity to study the organisation of long-term memory particularly since initial observations suggested sparing of episodic memory. More recent studies reveal, however, a more
complex but theoretically revealing pattern. On tests of autobiographical memory patients with semantic dementia show a "reverse step function" with sparing of recall of events from the most recent two to five years but impairment on more distant life periods. Anterograde recognition memory for visual materials is extremely well preserved, except in the most deteriorated cases, although performance is heavily reliant upon perceptual information about the studied stimuli, particularly for items which are no longer known by the subjects. On tests of verbal anterograde memory such as word learning, performance is typically poor even for words which are "known" to the patients. A source discrimination experiment, designed to evaluate familiarity - and recollection - based anterograde memory processes, found that patients with semantic dementia showed good item detection, although recollection of source was sometimes impaired. Semantic knowledge about studied items and measures of item detection and source discrimination were largely independent. The implications of these findings for models of long-term memory are discussed. The results support the concept that episodic memory, at least the recall of temporally specific autobiographical experiences, draws upon a number of separable memory processes some of which can function independently of semantic knowledge.

Authors: Nestor, P.J., GRAHAM, K.S., BOZEAT, S., Simons, J.S., HODGES, J.R.
Title: Memory consolidation and the hippocampus: Further evidence from studies of autobiographical memory in semantic dementia and frontal variant frontotemporal dementia
Reference: Neuropsychologia, 40 (6), 633-654
Year: 2002
Key Words: episodic memory, retrograde amnesia, semantic memory, temporal gradients, strategic retrieval, frontal lobes
Abstract:

Studies of autobiographical memory in semantic dementia have found relative preservation of memories for recent rather than remote events. As semantic dementia is associated with progressive atrophy to temporal neocortex, with early asymmetric sparing of the hippocampus, this neuropsychological pattern suggests that the hippocampal complex plays a role in the acquisition and retrieval of recent memories, but is not necessary for the recall of older episodic events. In an alternative view of memory consolidation, however, the hippocampus plays a role in the retrieval of all autobiographical memories, regardless of the age of the memory [71]. This 'multiple trace theory' predicts that patients with semantic dementia should show no effects of time in their autobiographical recall. In this article, we ask whether it is possible to reconcile the data from semantic dementia with the multiple trace theory by investigating whether the time-dependent pattern of autobiographical retrieval seen in the disease is due to (i) patients showing this effect being exceptional in their presentation and/or (ii) patients with semantic dementia exhibiting impaired strategic retrieval from concomitant frontal damage. A series of experiments in patients with semantic dementia, the frontal variant of frontotemporal dementia and Alzheimer's disease clearly demonstrates that neither of these two factors can explain the documented effect of time seen in semantic dementia. Nonetheless, we discuss how damage to semantic knowledge could result in an autobiographical memory deficit and suggest that data from semantic dementia may be consistent with both views of hippocampal involvement in long-term memory.

Authors: Simons, J., GRAHAM, K.S., HODGES, J.R.
Title: Perceptual and semantic contributions to episodic memory: Evidence from semantic dementia and Alzheimer's disease
Reference: Journal of Memory and Language, 47, 197-213
Year: 2002
Key Words: Recognition memory; Semantic memory; Episodic memory; Frontotemporal dementia;
Temporal lobe; Hippocampus; Perirhinal cortex

Abstract:

Previous group studies involving patients with semantic dementia, who have impaired semantic memory associated with temporal lobe atrophy, have documented the preservation of pictorial recognition memory. The present study replicated this general pattern, although analysis of patients' individual scores revealed evidence of recognition memory impairment in four of the patients with the most severe semantic deficit. Three factors that might contribute to this pattern of memory performance were examined: atrophic damage to medial temporal lobe regions, degradation of semantic representations, and disruption to visuoperceptual processes. Assessment of MRI scans revealed that atrophy affecting medial temporal lobe regions such as the perirhinal cortex accurately predicted recognition memory deficit. In line with this view, patients with early Alzheimer's disease, whose atrophy is thought to originate in this region, all showed severe recognition memory impairment. There was no evidence that the poor recognition memory seen at advanced stages of semantic dementia could be attributed directly either to degraded semantic knowledge or disrupted perceptual processing. Instead, the data suggested an impairment in the utilization of perceptual and semantic information in a lasting memory trace.

Authors: Garrard, P., Lambon-Ralph, M.A., HODGES, J.R.
Title: Semantic Dementia. A category-specific paradox.
Reference: Forde & Humphreys (Eds) Category-specificity in brain and mind, Psychology Press 149-179
Year: 2002
Key Words: Semantic dementia; temporal lobe; sensory-functional theory

Authors: GRAHAM, K.S., PATTERSON, K., Powis, J., DRAKE, J., HODGES, J.R.
Title: Multiple inputs to episodic memory: words tell another story. Episodic memory, semantic memory, semantic dementia, verbal recall, phonology
Reference: Neuropsychology 16(3), 380-389
Year: 2002
Key Words: episodic memory, semantic memory, semantic dementia, verbal recall, phonology
Abstract:

Seven patients with semantic dementia were given a memory task in which they were asked to recall and recognize 10-word lists of object-name vocabulary which had been pre-selected as either still ‘known’ — correct picture naming and word-picture matching — or now ‘unknown’ — incorrect picture naming and word-picture matching — to each individual patient. The patients showed a significant advantage for ‘known’ words in immediate free recall after several learning trials, and also in delayed recall and recognition. The majority of errors of commission for ‘known’ words were semantic, while phonological errors, especially blends of target words, were produced in the ‘unknown’ condition. These findings support claims (a) that episodic memory is supported by multiple inputs from semantic and perceptual systems, and (b) that success in verbal and non-verbal episodic memory tasks is differentially dependent upon semantic and perceptual information.

Authors: GRAHAM, K.S., KROPELNICKI, A., BIRD, H., Nestor, P., HODGES, J.R.
Title: Semantic dementia: a challenge to the multiple trace model of memory consolidation?
Reference: Eighth Annual Meeting of the Cognitive Neuroscience Society, 36
The fact that patients with bilateral hippocampal damage often show better recall of distant memories compared to more recent personal events has been cited as evidence that the hippocampal complex plays a early, and temporary, role in the retrieval of episodic memories. Further support for this view comes from studies of remote memory in patients with the neurodegenerative disease, semantic dementia, in which there is early sparing of the hippocampal complex in the context of focal atrophy to other temporal lobe regions. Patients with semantic dementia show better retrieval of recent autobiographical memories compared to those from the more distant past. An alternative theory proposes, however, that the hippocampus is important for the retrieval of all personal memories, regardless of their age. Further investigations of remote autobiographical memory in semantic dementia, and in patients with the frontal variant of fronto-temporal dementia, tested whether the pattern seen in semantic dementia was due to one or more of the following factors: (1) the reported patients are exceptional in their presentation; (2) impaired strategic retrieval due to concomitant frontal damage; and (3) linguistic deficits with accompanying preserved non-verbal access to autobiographical memory. These studies provide strong support for the view that the original pattern seen in semantic dementia is not artefactual.

Authors: Nestor, P.J., GRAHAM, K.S., HODGES, J.R.
Title: A reversed temporal gradient for episodic memory is seen in semantic dementia but not "frontal variant" frontotemporal dementia
Reference: Brain and Cognition, 47, 296-300

Studies of autobiographical memory in semantic dementia (SD, temporal variant of Frontotemporal Dementia) have revealed a "reversed" temporal gradient of impaired recollection; SD subjects show impaired recall of memories from distant relative to recent past. It has been proposed that this may be explained by impaired strategic retrieval secondary to concomitant frontal lobe damage. To test this hypothesis, a group of frontal variant Frontotemporal dementia (fvFTD) subjects (n=6 to 8) underwent two experiments using i/ the Autobiographical Memory Interview and ii/ a 20 item "free" Crovitz. In contrast to SD, fvFTD subjects showed no memory gradient although their absolute memory score on the Crovitz was significantly worse than controls. Implications for strategic retrieval, and, temporal and frontal lobe contributions to memory, are discussed.

Authors: ROGERS,T.T., Lambon Ralph, M.A., Plaut, D.C., HODGES, J.R., PATTERSON, K.
Title: Domain differences in semantic dementia: Implications for theories of category-specific deficits.

Semantic dementia (SD) is a syndrome marked by progressive and profound deterioration of semantic knowledge, consequent upon bilateral atrophy of the antero-lateral temporal lobes. This deficit is rarely characterized by significant category- or modality-specific effects: the great majority of SD patients are impaired for all semantic categories and all varieties of both input to and output from the semantic system, suggesting disruption to a unitary and amodal system of conceptual knowledge. Such patients do, however, show interesting differences in the kinds of errors they make in different semantic
domains. We have attempted to explain these differences with reference to a computational model in which central semantic representations emerge in a system that must learn the mappings among structured perceptual representations in different modalities. Domain effects arise as a consequence of differences in the degree to which objects in a given domain share structure in different modalities. We will consider the implications of this idea for theories of category-specific deficits, and will propose an extension of the unitary-semantics model that may account for different patterns of impairment in generalised semantic dementia vs category-specific cases.

Authors: GRAHAM, K.S., KROPELNICKI, A., Goldman, W.P., HODGES, J.R.
Title: Two Further investigations of autobiographical memory in semantic dementia
Year: 2003
Key Words: episodic memory, retrograde amnesia, semantic memory, temporal gradients, modality-specific effects
Abstract:

A number of investigations in semantic dementia have documented better retrieval of recent personal events compared to those in the more distant past (Graham and Hodges, 1997). Westmacott et al. (2001) challenged this result, however, finding relative preservation of remote memories in a single case of semantic dementia when he was tested using family photographs. In Experiment 1, we tested two possible explanations for the discrepancy between the published papers: (a) that there is a significant effect of modality in autobiographical retrieval in semantic dementia (e.g., patients will show better, or even preserved, remote memory when tested on nonverbal, compared to verbal, tasks); and (b) that the distinct pattern seen between patients is attributable to the different methods adopted for scoring the episodic quality of the memories. A patient with semantic dementia, AM, produced autobiographical memories to both words and family photographs. These personal events were scored by two raters using the scoring method described by Westmacott et al. (2001) and that reported by Graham and Hodges (1997). It was found that AM showed similar levels of remote memory impairment regardless of whether the cue was verbal or nonverbal. In addition, significant effects of time were revealed in AM’s performance on the verbal memory test, regardless of which method was used to score the memories. In Experiment 2, we investigated a related question: whether the relatively better retrieval of memories in semantic dementia could be due to over rehearsal of highly salient recent experiences? Four patients were tested on their ability to remember a recent event (either a trip to London or events from a Formula One Grand Prix race) using specially designed autobiographical questionnaires. It was found that all four were able to perform this task, although one patient showed evidence of some forgetting over three months, and another exhibited difficulty discriminating between two similar events. Together, the two experiments further confirm that patients with semantic dementia show a modality independent autobiographical memory deficit, with better retrieval of recent events.

Authors: Hovius, M., KELLENBACH, M.L., GRAHAM, K.S., HODGES, J.R., PATTERSON, K.,
Title: What does the Object Decision Task Measure? Reflections on the Basis of Evidence from Semantic Dementia
Reference: Neuropsychology, 17(1), 100-107
Year: 2003
Abstract:

The impact of semantic knowledge on visual object analysis was investigated by assessing the performance of patients with semantic dementia on an object matching test (Humphreys & Riddoch, 1984) and two object decision tests designed on different principles (Warrington and James, 1991b;
Riddoch and Humphreys, 1987a). On average, the patients achieved normal scores on both the object matching and the former object decision test, but were impaired on the latter object decision test, which was also the only visual object test that correlated significantly with degree of semantic impairment. These findings (a) demonstrate that object decision cannot be treated as a single task or ability and (b) suggest that it is not necessarily independent of conceptual knowledge.

Authors: HODGES, J.R.
Title: Semantic Dementia: A selective disorder of semantic memory?
Year: 2002

Authors: Lambon Ralph, M.A., PATTERSON, K., Garrard, P., HODGES, J.R.
Title: Semantic dementia with category specificity: A comparative case-series study
Reference: Cognitive Neuropsychology, 20(3-4-5-6), 307-326
Year: 2003
Abstract:

Patients with semantic dementia, the temporal variant of frontotemporal dementia, are relevant to both the neuroanatomical and neuropsychological debates in the category-specific literature. These patients present with a selective and progressive semantic deficit consequent on circumscribed atrophy of the inferolateral, polar temporal lobes bilaterally, including the inferotemporal gyrus. In this study, a patient KH with a significant advantage for artefacts over living things was compared to five other semantic dementia patients with commensurate levels of semantic impairment. KH demonstrated a consistent category-difference in favour of artefacts across all the expressive and receptive semantic tests. This difference was reliable even when familiarity, frequency and other potential confounding factors were controlled. While KH demonstrated an association between poor knowledge of sensory attributes and a consistently greater impairment on living things than artefacts, the other patients did not. As observed in a number of previous studies, all five of the patients, contrasted to KH, exhibited an advantage for functional/associative over sensory attributes but without demonstrating the category-specific deficit that the sensory-functional theory (and the locus of their atrophy) might predict. The results of this and other studies are discussed in relation to four accounts of category-specificity: the sensory-functions theory, domain-specific knowledge systems, intercorrelated features and individual differences.

Authors: HODGES, J.R., Miller, B.L.
Title: The neuropsychology of frontal variant FTD and semantic dementia: Introduction to the special topic papers: Part II
Reference: Neurocase, 7, 113-121
Year: 2001

Authors: GRAHAM, K.S.Murre, J.M.J., HODGES, J.R
Title: Episodic memory in semantic dementia: A computational approach based on the TraceLink model
Year: 1999
Authors: PATTERSON, K., HODGES, J.R
Title: Semantic dementia
Year: 2001

Authors: Davies, R., Xuereb, J.H., HODGES, J.R
Title: The human perirhinal cortex in semantic memory: An in vivo and post mortem MRI study in semantic dementia, Alzheimer's disease and matched controls
Reference: Neuropathology and Applied Neurobiology, 28(2)
Year: 2002
Key Words: Conference Proceedings & Published Abstracts
Abstract:

Introduction: Studies in macaque monkeys indicate that the perirhinal cortex is critical for processes analogous to human semantic memory (i.e. memory for words, objects, faces and concepts). It is selectively impaired in semantic dementia (SD), the temporal lobe variant of fronto-temporal dementia, and less consistently in Alzheimer's disease (AD). Materials and methods: In vivo volumetric magnetic resonance imaging (MRI) data were collected from 30 cases (10 SD, 10 AD, 10 controls). Regions were outlined on successive scan slices and volumes calculated. Hippocampus, temporopolar, perirhinal and entorhinal cortices were measured bilaterally (using the protocol of Insausti et al. Am J Neuroradiol; 19: 23). High-resolution MR images from six cadaver brains have also been obtained. These will further clarify the relationship between cytoarchitectonic boundaries and gyral/sulcal landmarks. Results: Initial analyses indicate greater atrophy of the perirhinal cortex in SD than in AD (P < 0.00002). There is also a trend towards greater atrophy of posterior than of anterior entorhinal cortex in AD vs. SD. Conclusions: Cortical regions of the medial temporal lobe, defined by reference to cytoarchitecture, have been measured on MRI. As predicted by primate studies, but for the first time in man, perirhinal cortex atrophy is correlated with a neuropsychologically defined syndrome of semantic memory impairment.

Authors: SIMONS, J.S., GRAHAM, K.S., PATTERSON, K., HODGES, J.R
Title: Semantic knowledge and episodic memory for famous faces in a case of semantic dementia
Reference: Proceedings of the British Psychological Society, 7, 45
Year: 1999
Key Words: Conference Proceedings & Published Abstracts

Authors: Thompson, S.A., PATTERSON, K., HODGES, J.R.
Title: Left/right asymmetry of atrophy in semantic dementia
Reference: Neurology, 61(9), 1196-1203
Year: 2003
Abstract:

Objective: To characterise presenting symptomatology in patients with semantic dementia (SD), and to investigate whether left and right temporal variants of the disease have distinct behavioral and cognitive profiles. Methods: Retrospective examination of case notes was performed in 47 consecutive referrals of patients with a clinical diagnosis of SD. Patients (and informants) were interviewed and underwent neuropsychological testing within six months of assessment. Patients were designated as
having predominantly left (L>R, n=36) or right (R>L, n=11) temporal lobe atrophy based on MRI or CT of the brain. Results: R>L and L>R SD groups did not differ in terms of presenting age (mean 63.4±7.4), symptom duration (3.6±2.3) or Mini-Mental State Examination scores (22.1±6.7). The most frequently reported behavioral symptoms were food fads, irritability, depression and preoccupation with puzzles; cognitive deficits included word-finding difficulties, reduced comprehension, difficulty with person identification and reduced conversation. The frequency of several symptoms differed significantly between groups. The following aspects were more likely to be associated with major right temporal atrophy: social awkwardness, job loss, loss of insight and difficulty with person identification. Word-finding difficulties and reduced comprehension were more salient features of L>R patients. Both groups showed deficits on semantic memory tests as expected for this syndrome, but several items yielded greater impairment for L>R than R>L. Conclusions: Patients with left and right predominant SD present with distinct behavioral-cognitive profiles. Characterisation of these features may assist in the early and accurate diagnosis of SD.

Title : Failing to get the gist: Reduced false recognition of semantic associates in semantic dementia
Reference : Neuropsychology (In Press)
Year : (In Press)
Abstract :

In two experiments involving patients with semantic dementia, the impact of semantic memory loss on both true and false recognition was investigated. The first experiment examined recognition memory for categories of everyday objects that shared a predominantly semantic relationship. There were significant differences between patients and controls in true and false recognition of objects from categories for which a large number of related exemplars were studied, but impairments diminished when category sizes were reduced. Consistent with these results, the patients showed preserved item-specific recollection for the pictorial stimuli, but compared with controls showed significantly reduced utilization of gist information regarding the categories of objects, a result consistent with the patients’ degraded semantic knowledge. To test this hypothesis, a second experiment used categories of abstract objects that were related to one another perceptually rather than semantically. Patients with semantic dementia showed no significant impairments in true or false recognition, and were indistinguishable from controls in terms of item-specific recollection and gist memory. Direct comparison between the two tasks confirmed a dissociation in semantic dementia between gist utilization for semantically-versus perceptually-related objects. These results suggest that the reduction in gist memory in semantic dementia is largely specific to semantic representations and cannot be attributed to general difficulty with abstracting and/or utilizing gist-like commonalities between stimuli.

Authors : BOZEAT, S., PATTERSON, K., HODGES, J.R.
Title : Relearning object use in semantic dementia
Reference : Neuropsychological Rehabilitation, 14(3), 351-363
Year : 2004
Key Words : Experimental, semantics, SD, praxis
Abstract :

The aim of this study was to investigate the ability of a patient with semantic dementia to re-learn to use objects that had, through the process of disease, become alien to her. Following only a brief demonstration of their use, JH achieved nearly perfect scores on the overall use of ten previously familiar objects. This dramatic boost in performance did decline steadily over time without any
practice. Even five weeks after the initial training, however, JH was still able to achieve an average score of around 60% which was significantly greater than at baseline assessment and sufficient to enable partially correct use of the objects. The results of this study demonstrate that it is possible for a patient in the advanced stages of semantic dementia, with severely degraded conceptual knowledge, to re-learn to manipulate objects through observation of their use. With continual reinforcement, in the form of more frequent training and practice sessions, it is likely that this improved performance could be maintained over time. A rehabilitation program based on this research may provide the rare opportunity for improvement in the face of this progressive disease: a glimmer of hope whilst many other cognitive functions are deteriorating.