Developmental amnesia: a new pattern of dissociation with intact episodic memory

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Abstract

A case of developmental amnesia is reported for a child, CL, of normal intelligence, who has intact episodic memory but impaired semantic memory for both semantic knowledge of facts and semantic knowledge of words, including general world knowledge, knowledge of word meanings and superordinate knowledge of words. In contrast to the deficits in semantic memory, there are no impairments in episodic memory for verbal or visual material, assessed by recall or recognition. Lexical decision was also intact, indicating impairment in semantic memory of vocabulary rather than absence of lexical representations. The case forms a double dissociation to the cases of Vargha-Khadem et al. [Science 277 (1997) 376; Episodic memory: new directions in research (2002) 153]; Gadian et al. [Brain 123 (2000) 499] for whom semantic memory was intact but episodic memory was impaired. This double dissociation suggests that semantic memory and episodic memory have the capacity to develop separately and supports models of modularity within memory development and a functional architecture for the developmental disorders within which there is residual normality rather than pervasive abnormality. Knowledge of arithmetical facts is also spared for CL, consistent with adult studies arguing for numeracy knowledge distinct from other semantics. Reading was characterised by difficulty with irregular words and homophones but intact reading of nonwords. CL has surface dyslexia with poor lexico-semantic reading skills but good phonological reading skills. The case was identified following screening from a population of normal schoolchildren suggesting that developmental amnesias may be more pervasive than has been recognised previously.

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1. Introduction

1.1. Residual normality

There is an on-going theoretical debate concerning the brain organisation of children with developmental disorders. Some argue that the organisation of cognitive systems in such cases may be fundamentally different from that of the normal child such that there is pervasive change to the system without residual normality (Thomas & Karmiloff-Smith, 2002). However, this view runs counter to many neuropsychological studies in which focal impairments have been described in children who appear to have intact residual skills (e.g. Anderson, Northam, Hendy, & Wrennall, 2001; Temple, 1997a,b). Moreover, the patterns of deficit and skill in such neuropsychological studies appear to integrate with models of normal development, suggesting impairments of components of a modular system (Clahsen & Temple, 2003; Temple & Clahsen, 2002).

Using such neuropsychological approaches, children who have specific reading difficulties despite normal intelligence are well documented and the form and character of these impairments has been systematically outlined in relation to the normal reading system (e.g. Castles & Coltheart, 1996; Jackson & Coltheart, 2001). Similarly, children who have developmental dysgraphias and developmental dyscalculias have been documented in detail and interpreted in relation to focal deficits with modular spelling and arithmetical systems (Rovet, Szkely, & Hockenberry, 1994; Sokol, Macaruso, & Gollan, 1994; Temple, 1985, 1991; Temple & Sherwood, 2002). Specific impairments in domains outside literacy have also been discussed. Developmental prosopagnosias in those of otherwise normal intelligence have been outlined (Ariel & Sadah, 1996; Campbell, 1992; Elgar & Campbell, 2001; Temple, 1992) as well as selective impairments in the perception of movement (Ahmed & Dutton, 1996) and selective impairments in the perception of location (McCloskey, Rapp, Yantis, & Rubin, 1995). In each of these cases, the
Disorders of memory in children were first described in detail over a decade ago, but it is only recently that there has been increased focus on them (Temple, 2002). The studies reported to date differentiate between acquired amnesias in children who have sustained injury or disease and developmental amnesias in which the impairment arises in the absence of an acquired lesion and any deterioration in skills. In practice, there is an issue about where, in this bipartite division, cases should be classified who have sustained injury or disease in utero or at birth. Such cases are acquired in the sense that an explicit aetiology is known but are developmental in the sense that the abnormality is present from birth and its impact emerges as the child acquires skills without any subsequent loss of these skills, however, the literature has not addressed this dichotomy and traditionally classifies the disorders one way or the other. The case of amnesia in a child following a prenatal stroke has been labelled developmental (Maurer, 1992; Temple, 2002), as have cases following birth anoxia (Vargha-Khadem, Gadian, & Connelly, 1997; Vargha-Khadem, Gadian, & Mishkin, 2001; Gadian et al., 2000; Baddeley et al., 2001).

1.2. Acquired versus developmental amnesias

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MS, a 22-year-old man reported by De Renzi and Lucchelli (1990), was born two months early with a birth weight of 250 g. EEG indicated bilateral bursts of theta and delta waves with a left frontotemporal propensity but there was otherwise no know neurological abnormality. MS complained of memory problems since childhood with difficulty in remembering the names and faces of familiar people, places, foreign language words, song lyrics, poems, mathematical formula and tables. Intelligence was normal, with a Verbal IQ of 110 and a Performance IQ of 111. Semantic memory for past events and famous names was impaired as was recognition memory for famous faces. Episodic memory was also impaired with difficulty in learning new verbal and nonverbal material, including story recall. Developmental amnesia with impairments of both episodic and semantic memory was also reported by Maurer (1992). NS had a CT scan when she was 9 years old that revealed low density areas in both temporal fossa indicating absence of the left temporal lobe and the pole and medial parts of the right temporal lobe. The aetiology, as mentioned above, was thought to be a prenatal stroke. Although both semantic memory and episodic memory were impaired, procedural memory was intact. Using simple stimulus-response training, NS was repeatedly trained to learn the names of people she met everyday. Results were effective as she was able to learn some names (Maurer, 1992).

Temple (1997a,b) reported a further case of development amnesia with both semantic memory impairment and episodic memory impairment. Julia was a 12-year-old girl for whom gestation appeared normal, and milestones were reached without significance. Memory difficulties were evident during pre-school years. At the age of six, Julia was diagnosed with temporal lobe epilepsy. There was impairment of semantic memory for factual knowledge and difficulties in episodic memory and the acquisition of new verbal and non-verbal material. Procedural knowledge, as assessed by recall of automated sequences like the alphabet and days of the week was normal.

1.3. Impairments of both semantic and episodic memory

Of those disorders classified as acquired amnesias of childhood, the first detailed case was that of Ostergaard (1987) who described a 10-year-old child, CC, who had sustained an anoxic episode with consequent damage to the left hippocampus. Semantic memory for facts was impaired, as was episodic memory, but procedural memory was intact. Acquired amnesia with impairment of both semantic and episodic memory was also described in a 9-year-old child, TC, following acute encephalopathy (Wood, Brown, & Felton, 1989). A further case with impairments of both episodic and semantic memory is reported by Broman et al. (1997). MS had severe asthma from an early age and suffered a respiratory arrest when eight years old. An MRI scan indicated loss of volume bilaterally in the medial temporal area, with some evidence of a shortened hippocampus. The authors suggested his profile closely resembled that of the adult case, HM (Corkin, 1984; Scoville & Milner, 1957).

In cases of developmental amnesia, in which there has been no known injury post-natally or at birth, impairments of both semantic memory and episodic memory have also been described. MS, a 22-year-old man reported by De Renzi and Lucchelli (1990), was born two months early with a birth weight of 250 g. EEG indicated bilateral bursts of theta and delta waves with a left frontotemporal propensity but there was otherwise no know neurological abnormality. MS complained of memory problems since childhood with difficulty in remembering the names and faces of familiar people, places, foreign language words, song lyrics, poems, mathematical formula and tables. Intelligence was normal, with a Verbal IQ of 110 and a Performance IQ of 111. Semantic memory for past events and famous names was impaired as was recognition memory for famous faces. Episodic memory was also impaired with difficulty in learning new verbal and nonverbal material, including story recall. Developmental amnesia with impairments of both episodic and semantic memory was also reported by Maurer (1992). NS had a CT scan when she was 9 years old that revealed low density areas in both temporal fossa indicating absence of the left temporal lobe and the pole and medial parts of the right temporal lobe. The aetiology, as mentioned above, was thought to be a prenatal stroke. Although both semantic memory and episodic memory were impaired, procedural memory was intact. Using simple stimulus-response training, NS was repeatedly trained to learn the names of people she met everyday. Results were effective as she was able to learn some names (Maurer, 1992).

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1.4. Dissociations in episodic and semantic memory impairment

The possibility of a dissociation between impairments of semantic and episodic memory in children emerged in the study of a 15-year-old girl, VT, who had amnesia following a mild head injury (Maravita, Spadoni & Parma, 1995). Although initially VT had impaired episodic and semantic memory, when reassessed 23 months later, episodic memory remained impaired but semantic memory had improved substantially. It appeared that she had partially recovered previously learnt material, as her learning of semantic knowledge was easier for material that she had been studying prior to her injury. Even so, her performance on semantic memory tasks never reached the previous level of proficiency and there remained a degree of impairment in both episodic and semantic domains (Maravita et al., 1995).
induces amnesia and these established skills usually remain intact. In children, language skills are still maturing. If the child is acquiring language with an impaired memory substrate, a semantic memory impairment could impinge on the development of the semantic representations that underpin language, resulting in impaired acquisition of vocabulary. Thus, in the child there are issues about the independence and interdependence of semantic systems which can be studied in a way that is not possible in adulthood.

Impairments in semantic knowledge of vocabulary in children have been described in cases where other semantic memory skills are also impaired. Thus, for example, in addition to impaired semantic memory for facts, CC had impaired memory for vocabulary (Ostergaard, 1987). MS who had impaired semantic knowledge of facts also had impaired semantic knowledge of words (Broman et al., 1997). Similarly, in addition to impaired semantic knowledge of facts, Julia had impaired semantic knowledge of words (Temple, 1997a,b). She had difficulties with both word recognition and word finding from the outset and prior to the onset of her epilepsy, Julia’s class teacher commented that she “sometimes has difficult remembering words or what a word actually means”. Language was characterized by an anomia such that at the age of 12.8 her naming age was 5.3. ON, described by Casalini et al. (1999) also had impaired memory for words and had difficulties in naming which affected her spontaneous speech. Whilst these may be coincidental co-existing impairments, it is also possible that there is a common underlying semantic impairment in each area of disorder. The semantic impairment which is the basis for impaired knowledge of facts may also underlie impaired knowledge of vocabulary. The only reported dissociation in development between semantic memory impairment for facts and semantic memory impairment for words is the case MS (De Renzi & Lucchelli, 1990). As discussed above, MS had impaired semantic memory for facts but semantic knowledge of vocabulary, for this 22-year-old man, was described as normal. There is comment in the study that there were difficulties with the names of places, people and certain foreign language words which means that the issue of a limitation in vocabulary knowledge cannot be dismissed completely but the case raises the possibility that semantic impairments for facts and vocabulary may be dissociable even in development. However, in the case reported here the semantic memory impairment for facts co-exists with impairment in semantic knowledge of vocabulary.

1.6. Impact of amnesia upon literacy in development

Contemporary models of reading identify a lexico-semantic reading route which utilises a semantic system of representations of words in order to subsequently activate their pronunciations (e.g. Coltheart, Rastle, Perry, Langdon, & Ziegler, 2001). Semantic impairment in reading would affect the development of this lexico-semantic reading route and would lead to the prediction of a particular pattern of developmental reading difficulty, namely surface dyslexia.
(Coltheart, Masterson, Byng, Prior, & Riddoch, 1983; Hanley, Hastie, & Kay, 1992; Temple, 1984, 1985, 1997a,b; Samuelsson, 2000). In developmental surface dyslexia, the impaired development of lexico-semantic reading systems leads to over-reliance on phonologically ("sounding out")-based reading systems. This leads to increased difficulty with irregular and exception words (e.g. pint, yacht) which violate rules of orthographic to phonological translation. There may also be an increased error rate with regular words because of the ambiguities of some of these rules in English but the difficulty with irregular words is the most marked. A further characteristic of reading in surface dyslexia is homophone confusion, where the meanings assigned to homophones (e.g. sale — sail) are confused. This is attributed to the over-reliance on the sound pattern of the words rather than their semantic representations, with meaning being derived after the phonological representation for the word. Connectionist models also predict surface dyslexia in association with semantic impairment (Plaut, McClelland, Seidenberg, & Patterson, 1996; Patterson et al., 1996). Surface dyslexia has been described in studies of adults with semantic dementia who have impairments of semantic memory (Patterson & Hodges, 1992; Patterson et al., 1996).

There has been no systematic published analysis of the pattern of reading skills in cases of semantic memory impairment in children, however, there are a number of reports of reading difficulty. CC had impaired semantic memory for facts and words but was also reported as having reading and spelling difficulties (Ostergaard, 1987). Similarly, MS had impaired semantic memory for facts and words and also had poor reading and spelling (Broman et al., 1997). MS had impaired semantic memory for facts and was also described as having reading and writing skills which were poorly developed for his age and were slow and laborious (De Renzi & Lucchelli, 1990). Our own case Julia (Temple, 1997a,b) in an unpublished investigation was found to be surface dyslexic. An early case of surface dyslexia was also reported in a child, NG, who had temporal lobe epilepsy and semantic memory impairment (Temple, 1984). These cases raise the issue of whether such an interrelationship might appear in other cases of semantic memory impairment in children. In the case reported here, impaired semantic memory extends to impairment in the lexico-semantic reading route and a pattern of surface dyslexia is demonstrated in reading.

1.7. Impact of amnesia upon arithmetical skills in development

Studies of memory impairments in adults suggest that general semantic knowledge about the world may dissociate from semantic knowledge about arithmetic. Cappelletti, Butterworth, and Kopelman (2001) and Butterworth, Cappelletti, and Kopelman (2002) have described a case of semantic dementia in a patient IH, a former banker, who had severely impaired semantic knowledge including the meanings of words and yet retention of calculation abilities, dependent upon knowledge of numerical facts. There was sparing of the ability to process the single semantic category of numbers. Knowledge of word meaning was severely impaired with inability to generate any items in category fluency tasks, inability to provide any accurate definitions of words, and severe anomia in spontaneous speech. In contrast, single and multi-digit addition and subtraction were excellent. Knowledge of multiplication tables though impaired in comparison to controls was nevertheless 75% accurate, demonstrating a markedly higher level of ability than seen in vocabulary tasks. In literacy, there was severe impairment in reading and writing non number words but retained ability to write number words (Cappelletti, Kopelman, & Butterworth, 2002). IH could write three but not tree, and seventeen but not event. These literacy skills form a double dissociation with the developmental case, Paul (Temple, 1989), who could read and write non number words well but had digit dyslexia. Paul could read orchestra, colonel and physics but made errors reading three, nine and five. The study of IH (Butterworth et al., 2002; Cappelletti et al., 2001, 2002) raises the further issue of whether semantic memory impairment in children would always extend to encompass arithmetical semantic knowledge or whether general semantic knowledge about the world and arithmetical semantic knowledge may dissociate in development. Temple (1994) described twins with significantly impaired semantic knowledge of numerical facts who had excellent general semantic knowledge about the world and good vocabulary knowledge. The reverse dissociation has not previously been described but is demonstrated in the case reported here.

1.8. Intact intelligence

The cases of acquired amnesia in children indicate limitations in the impact of the memory impairment upon other domains. Broman et al. (1997) argued that despite his amnesia, MS had syntactic and logical reasoning skills which had matured to adult proficiency. TC (Wood et al., 1989; Ostergaard & Squire, 1990) also made scholastic progress, albeit limited. Some of the current studies suggest that acquired amnesia may be compatible with normal intelligence (Vargha-Khadem et al., 2002). The developmental case MS (De Renzi & Lucchelli, 1990) also had good normal intelligence on formal psychometric assessment. If developmental amnesia can co-exist with normal intelligence then there may be developmental amnesias within the normal school population.

It is perhaps surprising that a larger number of cases of developmental amnesia have not been reported. One possibility is that the developmental amnesias are rare and that the reported cases reflect the incidence. Another possibility is that there are children with developmental amnesia whose cases have not been identified. In pursuit of this issue, the current study involved screening normal children attending normal schools to determine whether within this population,
2. Method

2.1. Screened participants

The screened participants were 239 8–9-year-old children recruited from mainstream primary schools with mixed catchment areas in Colchester, Essex and the surrounding district. Criteria for inclusion consisted of absence of known learning disability and absence of psychotropic medication, as specified by parents. Parental consent was obtained. Children were administered the screening battery at their schools, with group testing within classes.

2.2. Screening battery

A screening battery was constructed to provide an estimate of general intelligence; a measure of semantic memory for established facts; and a measure of episodic memory for recently encoded information.

2.2.1. Intelligence

Raven’s Standard Progressive Matrices (Raven, 1999) is a multiple-choice non-verbal intelligence test. Participants have to decide which amongst distracters is the target which forms the appropriate match to a given pattern or set of patterns. For brevity, only alternating items of sets A–E were used, and a pro rata score was deduced.

2.2.2. Semantic memory screener

Information is a verbal subtest from the Wechsler Intelligence Scales for Children (WISC-IIIUK) (Wechsler, 1992). It consists of a series of questions regarding general knowledge that are read aloud. It therefore tests semantic memory for established factual knowledge. For its use as a screening test, participants had to write down their answers, rather than saying them aloud which would have been the standard procedure.

2.2.3. Episodic memory screener

Episodic memory was assessed using a modification of the Rey Auditory-Verbal Learning Test (Rey, 1964). Participants were required to listen as a list of 20 words taken from the Rey lists were read aloud, after which they had to write down, rather than say aloud, as many as they could remember.

2.3. Screening results

The average IQ of the 239 children tested, as assessed by Raven’s Matrices was 108.74 (standard deviation (S.D.) = 14.80). The average age-scaled score for semantic memory, as assessed by the Information subtest of the WISC-IIIUK, was 10.49 (S.D. = 3.19). The average episodic memory score, as assessed by the modified Rey was 7.88 (S.D. = 2.51).

It was evident that amongst the children screened, there were some who were of normal, average intelligence but who had impaired scores on episodic and/or semantic memory. In the sample of children assessed here there were 134 who had average intelligence, defined strictly as within one standard deviation of the group mean. Of these 2 had weak scores on semantic memory but not episodic memory, 3 had weak scores on episodic memory but not semantic memory, and three had weak scores on both episodic and semantic memory. Thus the incidence of memory impairments in those of average intelligence was 8/134 (5.9%). In a linked study of 70, 11–12-year-old children from normal schools, 52 were found to have average IQs using the same strict criterion and of these 3 had weak episodic and/or semantic memory, an incidence of 3/52 (5.8%).

One of the 8–9-year-old children, CL, who had average intelligence and had weak scores on episodic memory but not semantic memory on the screening measures was available for further study and is the focus of the current paper. A second 8–9-year-old child MM, who had normal intelligence but weak scores on both the episodic memory and semantic
memory screeners was also studied in detail and is reported in a separate paper (Temple & Richardson, forthcoming).

2.4. Memory study participants

CL had a normal IQ of 115. She also scored normally on the test of episodic memory with a score of 7. However on the test of semantic memory she attained an age-scaled score of only 5. CL was selected for further detailed investigation as a possible case of developmental amnesia with impaired semantic memory but intact episodic memory.

The control group was selected in order to compare CL’s memory skills with those of a group of children of average intelligence, matched for age who had normal memory scores. The control group was therefore selected from the screened children on the basis of having intelligence scores, semantic memory scores and episodic memory scores all of which were within one standard deviation of the mean score of the screened children. The control group was thus a group of typically developing children with average intelligence and memory scores on the screening measures. All of the controls were reported by parents to have reached developmental milestones within normal limits. Parents reported no significant difficulties with school in relation to reading, writing or memory. None had extra classroom assistance. The control group consisted of eleven of the children who fitted these criteria and whose parents agreed that they might take part in the more detailed and time-consuming study. There were six females and five males.

2.5. CL case history

CL was 9 years old at time of investigation. She has no siblings. Pregnancy was carried to full-term with no evident difficulties. CL’s medical notes report a normal neonatal examination and normal 6-week examination. No specific help was required in the neonatal period. Developmental milestones were uneventful. CL began walking at around 9–12 months, and she began babbling around 12–15 months. CL attends a mainstream primary school. No major problems at school were reported. CL herself reported that she enjoyed most of her classes, though she stated that English was not her best subject.

3. Protocol of memory tasks

3.1. Semantic knowledge of facts

3.1.1. General knowledge for facts

The participant was asked a series of general knowledge questions, adapted from the Trivia Quiz book (Henderson Publishing, 1995). This tested recall of established memories for facts. If the answer was not known or an incorrect answer was given, then the participant was presented with a choice of three answers, consisting of the correct answer and two distractors from which a forced choice decision was to be made. For example one question was “What is the name of the flag on a Pirate ship?” The alternatives were A: Blue Peter, B: Jolly Roger (correct), C: Happy Dave. The maximum possible score was 75.

3.1.2. Arithmetical facts

Using a procedure similar to Warrington (1982) and Temple (1991), the digits 2, 3 and 5 were paired with the numbers 1–10, generating 30 digit pairs. Each of the pairs was randomly presented orally to participants as an addition, subtraction and multiplication question. For division, the digits 2, 3 and 5 were paired with those numbers 1–10 that could be divided exactly by these digits; this generated 10 division questions. The maximum possible score was therefore 30 each for addition, subtraction and multiplication and 10 for division, a total of 100.

3.2. Semantic knowledge of words

3.2.1. Lexical decision

The Spot-the-Word test is a task from the Speed and Comprehension of Language Processing Test (SCOLP) (Baddeley, Emslie, & Nimmo-Smith, 1993). Participants were asked to decide, for each verbally presented pair, which was a real word, the distracter being a nonword. This task determines knowledge of words names but need not require semantic knowledge of the words. This was an untimed task of ability to access previously stored lexical representations. The maximum possible score was 40.

3.2.2. Receptive vocabulary

Receptive vocabulary was assessed using a subtest of the Psycholinguistic Assessment of Language Processing and Aphasia (PALPA) (Kay, Lesser, & Coltheart, 1992). Participants were shown a booklet of 40 pages, each page containing five pictures. They had to point to the picture, matching the word spoken by the experimenter, the other four pictures being distracters that were closely or distantly semantically related to the target. This task requires semantic knowledge about words. The maximum possible score was 40.

3.2.3. Conceptual organisation

This task was based on a questionnaire designed by Laiacina, Barbarotto, & Capitani (1993) in which participants had to answer various questions about pictures they had previously seen. Each question requires semantic knowledge about the word. Items were based on the PALPA (Kay et al., 1992) and were divided into six categories of which three were living things: animals, fruit and vegetables, and three were non-living things: tools, furniture and vehicles. The 240 questions, six for each item, were presented orally in a standardised random order. Firstly, the name of the item was provided and then the participants were asked the six questions in the same order. Examples are as follows:
• Subordinate: associative functional features for the item bicycle: Does it carry one person, about five persons or about fifty persons?
• Subordinate: associative contextual features for the item apple: Does it grow on a tree, on the ground or on a bush?

The participants were required to select the correct response from the three orally presented alternatives. The maximum possible score was 240.

3.2.4. Semantic verification

This task is from the SCOLP (Baddeley et al., 1993). Participants judged the veracity of verbally presented statements that were either factually correct or nonsense items, for example “Figs are people.” “Lettuce can be eaten.” These judgements require semantic knowledge about words. Responses were recorded for accuracy within a 120 s time limit. There were 50 items in total.

3.3. Episodic memory

3.3.1. Visual recall

The Design Memory subtest from the Wide Ranging Assessment of Memory and Learning (WRAML) (Sheslow & Adams, 1990) is a measure of visual memory. Participants were shown a card portraying a line drawing of a design for five seconds. The card was then removed and after waiting for 10 s, the participants had to reproduce the design on a piece of paper with a pencil. This procedure was repeated with four separate cards. The maximum possible score was 55.

3.3.2. Visual recognition

In the Picture Memory subtest from the WRAML (Sheslow & Adams, 1990), participants were shown a card depicting a scene, e.g. the seaside for 10 s, and then shown a similar card with minor alterations. Participants had to identify the differences. This was repeated with four separate cards. The maximum possible score was 47.

3.3.3. Verbal recall

A verbal counterpart to the Design Memory subtest is the Story Memory subtest from the WRAML (Sheslow & Adams, 1990). The participants were read a short story, lasting about 60 s, and then asked to repeat back as much as they could remember, trying to use the same phrases as given in the story. The maximum possible score was 51.

3.3.4. Verbal recognition

The Story Memory Recognition subtest from the WRAML (Sheslow & Adams, 1990) includes an extension of the previous test. Immediately after recalling the story, participants were asked a series of multiple-choice questions regarding specific details contained within the text. The maximum possible score was 30.

3.3.5. Warrington recognition test

The Recognition Memory Test (Warrington, 1984) provides an assessment of recognition memory for verbal material (words) and nonverbal material (faces). The same procedure was used for both formats. Participants were presented with 50 stimulus items at a rate of approximately one every 3 s, and were required to state whether or not the stimulus was pleasant or unpleasant. Recognition was then tested in a forced choice procedure; pairs were composed of one original stimulus coupled with a distracter stimulus with participants required to indicate which of the two they had seen before. The maximum possible score for words was 50 and the maximum possible score for faces was 50.

3.4. Autobiographical memory

The Autobiographical Memory Interview (AMI) (Kopelman, Wilson, & Baddeley, 1990) is a semi-structured interview that objectively measures the personal semantic and autobiographical knowledge of participants. The first component assesses the recall of facts from their own history, e.g. “What was the name of your first school?” The second component assesses recall of specific events and incidents from their own history, e.g. “Tell me about your first day at school.” Each component of the AMI assesses personal memories across three broad time bands: childhood, early adult life and recent events.

The Childhood section of the AMI test was given, which addresses two stages of childhood: Infancy 0–5 years and later childhood 5–11 years. In order to prevent a ceiling effect on scores, the AMI was modified with additional questions being added to both stages. The total potential scores for personal semantic and autobiographical questions were raised from 13 to 24 and 6 to 18 respectively. Additional questions in the personal semantic sections were, for example, place of first holiday, name of nursery school (if any), first film seen at the cinema, first book you read alone. Example of additional questions in the autobiographical sections were first memory of Christmas, first day at school, first holiday. The categorisation of the additional questions to the appropriate age band 0–5 years or 5–11 years was probabilistic rather than absolute. For example, first film seen at the cinema was added to the Sections 5–11 years even though some children might go to the cinema at a younger age. Veracity of answers was corroborated against those of parents/guardians. The total possible score was 42.
3.5. Procedural memory

The Mirror-Drawing Task (Milner, 1965) consisted of instructing the participants to draw a line within the boundaries of a star-shaped target, i.e. a star superimposed onto a larger star but without seeing this directly. Instead, they had to draw the line via the star's reflection in a mirror. Participants attempted this over two test sessions, with a total of six trials given, three per session. Error scores were derived from the number of times the pen crossed the inner or outer line of the star-shaped target.

3.6. Learning with repeated presentations

3.6.1. Visual memory

Visual Learning is a WRAML subtest that required the participants to learn where designs were located on a board (Sheslow & Adams, 1990). Before the test began, they were shown the location of 14 coloured designs on the board, hidden under foam covers. They were then shown pictures of a design and asked to point to its location over four trials. If the answer was correct, the foam cover was lifted; if the answer was wrong, then only the true location was revealed. The maximum possible score per trial was 14.

3.6.2. Verbal memory

The Selective Reminding Memory Test is an adaptation of the Rey Auditory-Verbal Learning test using the complete list of words (Rey, 1964). The participants recalled as many words as they could in any order, from a list of 15 previously read out. After each trial, the examiner repeated only the words that the participants omitted in that trial; there were five trials. The maximum score per trial was 15.

3.6.3. Visual novel-paired associates

This test is from the Wechsler Memory Scale-R (WMS-R) (Wechsler, 1987). Participants were shown six abstract line drawings, each uniquely paired with a colour. They were then shown the line drawings individually and asked to indicate the corresponding colour associated with each figure. This procedure was repeated six times. The maximum score per trial was six.

3.6.4. Verbal novel-paired associates

This test is also from the WMS-R (Wechsler, 1987). Participants were read a group of eight word pairs, then were read the first word of each pair, and required to supply the second word from memory. This was repeated six times. Four pairs consisted of words for which there was an established semantic relationship, whilst four pairs contained unrelated words. The maximum score per trial was eight.

3.7. Behavioural memory

The Rivermead Behavioural Memory Test (Wilson, Cockburn, & Baddeley, 1985) contains a series of tests of memory and recall ability. It measures the everyday consequences of memory difficulties as could be encountered in a real-life context.

4. Protocol of literacy tasks

4.1. Reading

4.1.1. Single words

The Schonell Graded Word Reading Test (Schonell & Schonell, 1956) was administered to participants to measure single word reading skill. The test involves reading a list of single words aloud. Total score was 100.

4.1.2. Text

The Neale Analysis of Reading Ability Test (Neale, 1989) measures three facets of text reading: accuracy, speed and comprehension. Using Form 1, participants were timed reading short stories and were marked on accuracy of reading. The story they had just read was then removed and they had to answer questions about it.

4.1.3. Nonwords

Participants were presented with the nonwords from the Graded Nonword Reading Test (Snowling, Stothard, & McLean, 1996). They were asked to read each one aloud. Total score was 31.

4.1.4. Regular and irregular words

The stimulus list of Behrmann & Bub (1992) was used. This consists of two lists of 60 words, one containing regular and the other irregular words, in terms of spelling-to-sound patterns. In each set of sixty words, there were six frequency bands, each containing 10 words. The participants had to read the words presented in random order and were marked for accuracy. Total score for regular words was 60 and for irregular words was 60.

4.1.5. Homophones

Participants had to read a list of 40 words containing 20 homophone pairs. They were then asked what each word meant (Temple, 1984). Total score was 40.

4.1.6. Morphological forms

Participants had to read Marshall’s Morphological List (unpublished), which contained both base lexical items and those with derived or inflectional form. Responses were marked for accuracy. There were 96 words in total.

4.2. Spelling

4.2.1. Single words

The Schonell Graded Word Spelling Test (Schonell & Schonell, 1956) was administered to participants to measure
spelling skill. Single words were dictated aloud for written spelling. Total possible score was 100.

4.2.2. Regular and irregular words
The stimulus list of Behrmann & Bub (1992), as described above, was dictated to the participants in random order for written spelling. Total score was 60 for regular words and 60 for irregular words.

4.2.3. Nonwords
Participants were read a list of nonwords (Snowling et al., 1996). They were instructed to write each word as it sounded. Total score was 31.

4.2.4. Homophones
Participants were read a list of 40 words, which contained 20 homophonic pairs, and their definitions. They had to write them down (Temple, 1984). Total score was 40.

5. Screening results
CL was compared to controls using z-tests. Table 1 compares the performance of CL to the control group on the screening measures. There were no significant differences between CL and the control group in age, IQ or episodic recall and performance was also within the control range on all three measures. Performance on the screening test for semantic memory was significantly lower for CL than for controls (z = 4.38, P < 0.001) and CL’s score was also below the control range.

6. Memory results
For each task, CL’s performance was classified as impaired in relation to controls if it differed significantly from controls using z-tests of statistical significance or if it was below the control range and worse than any of the controls. Given that the exploration was of memory impairment and the a priori expectation that where there was a difference, the predicted direction would be of inferior performance by CL, one-tailed test were used throughout.

6.1. Semantic knowledge of facts
The means and standard deviations (S.D.s) for results in this section are given in Table 2.

Table 1
Screening results for CL and the selected controls

<table>
<thead>
<tr>
<th>Test</th>
<th>CL</th>
<th>Controls</th>
<th>Min</th>
<th>Max</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>8.98</td>
<td>8.33</td>
<td>9.58</td>
<td></td>
</tr>
<tr>
<td>Semantic memory screener</td>
<td>10.64</td>
<td>9.06</td>
<td>11.15</td>
<td></td>
</tr>
<tr>
<td>Semantic memory screener</td>
<td>10.64</td>
<td>9.06</td>
<td>11.15</td>
<td></td>
</tr>
<tr>
<td>Episodic memory screener</td>
<td>8.15</td>
<td>6.55</td>
<td>10.60</td>
<td></td>
</tr>
<tr>
<td>IQ</td>
<td>115</td>
<td>105.45</td>
<td>115</td>
<td></td>
</tr>
</tbody>
</table>

* Statistically significant, see text.

6.1.1. General knowledge for facts
There are three sets of results indicating performance on free recall of answers, forced-choice recall of answers and the total correct by both means. On free recall of answers, CL is significantly poorer than the controls (z = 1.68, P < 0.05) and also below the control range. On forced choice recall of answers to a question answered incorrectly, the difference between CL and controls is not significant (z = 1.35, P > 0.09). On the combined total for free recall and forced-choice recall out of a possible 75, CL is significantly poorer than controls (z = 2.84, P < 0.002) and below the control range.

6.1.2. Arithmetical facts
The scores are number correct out of 100. Performance for CL and controls was close to ceiling on these measures with performances of high accuracy. CL demonstrated no impairment in relation to controls.

6.2. Semantic knowledge of words
6.2.1. Lexical decision task
The scores indicate how many words, out of 40 pairings of words and nonwords, were identified correctly. CL performed at a normal level for age and was not significantly impaired on this task of word recognition.

6.2.2. Receptive vocabulary
The scores indicate the number of correctly identified target picture in response to the spoken words. CL was significantly impaired in comparison to the controls (z = 2.24, P < 0.02).

6.2.3. Conceptual organisation
The results for this test are given in Fig. 1 and for the Superordinate category in Table 1. The figures show the correct scores obtained for each level of categorical organisation. Performance averaged across the categories revealed...
no significant differences between CL and controls (34 versus 35.43 respectively; \( z = 0.45, P > 0.33 \)), but scores for the Superordinate Category level were significantly lower for CL than for controls (\( z = 4.25, P < 0.001 \)).

6.2.4. Semantic verification
The scores indicate how many statements were correctly identified as either making sense or being nonsensical within the time limit of 2 min. There was no significant difference between CL and controls (\( z = 1.21, P > 0.11 \)).

There were multiple comparisons in this section, with 8 across the tasks whose data is presented in Table 2, and 6 across the tasks whose data is presented in Fig. 1. One significant result might have been expected by chance, however, there were four significant results.

6.3. Summary of semantic knowledge
The results indicate significantly impaired knowledge of semantic facts, confirming the impairment detected on the screening measure of semantic knowledge. In relation to words, CL was able to recognise word names, indicating that a store of vocabulary was established, but for some tasks requiring semantic knowledge of these words, namely word comprehension and conceptual judgements about words at the superordinate level she was significantly impaired.

6.4. Episodic memory

6.4.1. Visual and verbal recall & recognition
The results are given in Table 3. The scores refer to the number of items correctly recalled and recognised. CL did not differ significantly from controls on any of the six episodic memory tasks. Her performance was also within the control range on all six episodic memory tasks. There was therefore no evidence of any episodic memory impairment.

<table>
<thead>
<tr>
<th>Test</th>
<th>CL</th>
<th>Controls</th>
<th>Min</th>
<th>Max</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visual Recall</td>
<td>41</td>
<td>34.56 (7.00)</td>
<td>24</td>
<td>45</td>
</tr>
<tr>
<td>Visual Recognition</td>
<td>23</td>
<td>23.36 (5.18)</td>
<td>15</td>
<td>30</td>
</tr>
<tr>
<td>Verbal Recall</td>
<td>26</td>
<td>31.6 (7.11)</td>
<td>17</td>
<td>41</td>
</tr>
<tr>
<td>Verbal Recognition</td>
<td>23</td>
<td>24.2 (2.04)</td>
<td>22</td>
<td>28</td>
</tr>
<tr>
<td>Warrington recognition</td>
<td>36</td>
<td>37.82 (4.12)</td>
<td>32</td>
<td>43</td>
</tr>
<tr>
<td>Words</td>
<td>32</td>
<td>35 (5.61)</td>
<td>29</td>
<td>42</td>
</tr>
</tbody>
</table>

Note, the potential total scores for the recall and recognition conditions differ.

6.5. Autobiographical memory
The results are given in the Fig. 2. The scores refer to number of answers judged to be correct by parents/guardians. CL did not differ significantly from controls in performance on personal semantic questions nor on episodic-type autobiographical memories. Thus, CL’s autobiographical memory did not differ from normal.

<table>
<thead>
<tr>
<th>Memory (by age)</th>
<th>CL</th>
<th>Controls</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sm (0-5)</td>
<td>I</td>
<td>I</td>
</tr>
<tr>
<td>Aut (0-5)</td>
<td>I</td>
<td>I</td>
</tr>
<tr>
<td>Sm (5-12)</td>
<td>I</td>
<td>I</td>
</tr>
<tr>
<td>Aut (5-12)</td>
<td>I</td>
<td>I</td>
</tr>
</tbody>
</table>
6.6. Procedural memory

The results for trace-a-star task are given in Fig. 3. The scores refer to the number of errors made, i.e. transgressing the outer or inner line of the tracing star. A lower score indicates better performance. CL initially exhibited a learning trend and made fewer errors over consecutive trials, from trials 1–2 and 3–4. However, she then reversed this trend on trials 4–6 and made more errors. Thus, although looking initially similar to controls the increase in error rate over later trials meant that by the final trial CL’s performance was significantly different from controls ($z = 1.81, P < 0.04$). On this task, the pattern of procedural memory is abnormal and performance is significantly impaired.

6.7. Learning with repeated presentations

6.7.1. Visual memory and verbal memory

The results for this test are given in Fig. 4a and b. The scores refer to the number of items correctly recalled. CL exhibited a learning curve in both visual and verbal modalities, cumulatively acquiring more correct responses over each successive trial. On visual items, performance across the four trials did not differ significantly from controls ($z = 1.29, P > 0.10$), though delayed visual recall was poorer than controls ($z = 1.75, P < 0.04$). On verbal items, the initial trial which required episodic recall did not differ significantly from controls ($z = 1.11, P > 0.05$), consistent with the absence of impairment on the screening test for episodic memory and the absence of impairment on the other episodic memory measures detailed above. Mean performance across verbal trials was not significantly poorer than controls, nor was delayed verbal recall.

6.7.2. Novel-paired associates

The results for these tasks are given in Fig. 5a and b. The scores refer to the number of items correctly recalled. For visual paired associates, CL attained ceiling performance from the first trial, and for verbal paired associates CL attained ceiling performance from the second trial. Both sets of scores indicate normal paired-associate learning.

6.8. Summary of learning with repeated presentations

Performance on all tasks with repeated presentations was normal in comparison to controls. However delayed recall scores were poorer than normal on visual learning.
6.9. Behavioural memory

Across the subtests of the Rivermead Behavioural Memory Test the total possible score is 22. CL attained a score of 21 compared to a mean control score of 20.7 (S.D. = 1.64). CL’s performance did not differ significantly from controls.

7. Literacy results

7.1. Reading

The means and S.D.s for the results of this section are given in Table 4.

7.1.1. Single words and text

The scores refer to the number of words read correctly for the single words on the Schonell and the text reading ages on the Neale. CL’s ability to read single words on the Schonell was significantly poorer than controls (z = 1.77, P < 0.05). Accuracy of text reading did not differ significantly from controls, nor did comprehension of text but rate of reading text was significantly slower than controls (z = 2.71, P < 0.003).

7.1.2. Nonwords

The score refers to the number of nonwords read correctly out of 31. CL’s ability to read non-words did not differ significantly from controls (z = 1.28, P > 0.10) and was also below the control range.

7.1.3. Regular and irregular words

The results are given in Fig. 6a and b. Scores refers to the number of words out of 10 read correctly. CL displayed a frequency effect, scoring more highly the more common words became. Her performance was significantly poorer than controls for regular words across all frequencies (z = 3.29, P < 0.001). On irregular words, performance was significantly poorer on the 10–19, 50–99 and 100–199 m frequencies (z = 2.22, 2.16 and 3.02 respectively, all P < 0.02). There was a regularity effect across the three lowest frequency bands (χ² = 6.67, P < 0.01, with Yates correction for continuity).

7.1.4. Homophones and morphological forms

CL made significantly more homophonic errors than controls (z = 2.22, P < 0.02) and was outside the control range.

Table 4

Results for reading

<table>
<thead>
<tr>
<th>Test</th>
<th>CL</th>
<th>Controls</th>
<th>Min</th>
<th>Max</th>
</tr>
</thead>
<tbody>
<tr>
<td>Single words</td>
<td>35*</td>
<td>60.55 (14.39)</td>
<td>34</td>
<td>78</td>
</tr>
<tr>
<td>Text Accuracy</td>
<td>8.33</td>
<td>10.08 (1.52)</td>
<td>8.17</td>
<td>12.07</td>
</tr>
<tr>
<td>Rate</td>
<td>6.5*</td>
<td>10.37 (1.43)</td>
<td>8.08</td>
<td>13</td>
</tr>
<tr>
<td>Comprehension</td>
<td>9.91</td>
<td>9.58 (1.28)</td>
<td>7.94</td>
<td>11.92</td>
</tr>
<tr>
<td>Non-Words</td>
<td>18</td>
<td>21.64 (2.84)</td>
<td>16</td>
<td>25</td>
</tr>
<tr>
<td>Homophonic errors</td>
<td>14*</td>
<td>4 (3.46)</td>
<td>0</td>
<td>10</td>
</tr>
<tr>
<td>Morphological Forms</td>
<td>83</td>
<td>89.27 (4.47)</td>
<td>82</td>
<td>95</td>
</tr>
<tr>
<td>Receptive vocabulary written</td>
<td>39</td>
<td>39.18 (0.60)</td>
<td>38</td>
<td>40</td>
</tr>
</tbody>
</table>

*a Statistically significant, see text.

![Fig. 6. (a) Results for reading regular words. (b) Results for reading irregular words.](image-url)
range. She did not differ significantly from controls on reading morphological forms ($z = 1.40, P > 0.08$).

7.2. Reading skills summary

CL was significantly impaired in word recognition skills in comparison to controls. There was normal reading of nonwords, impaired reading of irregular words, a regularity effect at low frequencies and homophone confusion. CL displayed the classical characteristics of developmental surface dyslexia.

7.3. Spelling

The means and S.D.s for the results of this section are given in Table 5.

7.3.1. Single words

The scores refer to the number of words spelt correctly. CL was significantly impaired in comparison to controls ($z = 1.69, P < 0.05$) and performed below the control range.

7.3.2. Nonwords

The score refers to the number of non-words out of 31 spelt appropriately. CL was not significantly impaired in comparison to controls ($z = 0.15, P > 0.44$).

7.3.3. Regular and irregular words

The results are given in Fig. 6a and b. The scores refer to the number of words spelt correctly in each frequency band out of 10. CL showed a frequency effect, spelling regular words of high frequency more easily than those of low frequency. Her performance was significantly worse than controls at spelling regular words averaged across all frequencies ($z = 1.70, P < 0.05$); though her ability to spell irregular words did not differ from controls when averaged across the frequencies ($z = 1.09, P = 0.14$). There was no effect of regularity ($\chi^2 = 0.04, P > 0.84$ with Yates correction for continuity).

7.3.4. Homophones

The results are given in Table 5. The scores refer to the number of words misspelt with homophonic errors. CL made significantly more homophonic errors than controls ($z = 3.65, P < 0.001$) and the number of errors was outside the control range.

Table 5

<table>
<thead>
<tr>
<th>Test</th>
<th>CL</th>
<th>Controls</th>
<th>Min</th>
<th>Max</th>
</tr>
</thead>
<tbody>
<tr>
<td>Single Words</td>
<td>13$^*$</td>
<td>56 (33.57)</td>
<td>34</td>
<td>74</td>
</tr>
<tr>
<td>Nonwords</td>
<td>25</td>
<td>25.45 (3.01)</td>
<td>20</td>
<td>30</td>
</tr>
<tr>
<td>Homophonic errors</td>
<td>15$^*$</td>
<td>4.67 (2.83)</td>
<td>1</td>
<td>10</td>
</tr>
</tbody>
</table>

*Statistically significant, see text.

7.3.5. Homophones

The results are given in Table 5. The scores refer to the number of words misspelt with homophonic errors. CL made significantly more homophonic errors than controls ($z = 3.65, P < 0.001$) and the number of errors was outside the control range.

Fig. 7: (a) Results for spelling regular words. (b) Results for spelling irregular words.
7.4. Spelling skills summary

Spelling of non-words was not impaired indicating in-tact phonological spelling skills. Spelling of both regular and irregular words was impaired. Spelling of homophones was also impaired. These indicate impaired lexico-semantic spelling systems Fig. 7.

8. Discussion

8.1. Developmental amnesia

Until relatively recently, the very existence of any form of developmental amnesia was in doubt. The assertion that a focal pattern of impairment in developmental memory could not be observed in isolation was traditionally explained by two contrasting theories: that plasticity and adaptability in the developing brain compensates for loss of function (e.g. Kennard, 1936, 1938); and the immature system’s adaptation without a pre-specified functional architecture, creates pervasive and global impairments to the cognitive system that may bury any specific memory impairment (e.g. Hebb, 1942). Nevertheless, there have now been a few case descriptions of developmental dyssnesia. This raised the issue of whether these rare reports simply represented a rare occurrence or limited identification of a more common disorder. One objective of the current study was to determine whether developmental amnesia existed within the normal population of school children. By screening a large sample of children from mainstream schools, a number of children of normal intelligence with weak semantic and/or episodic memory skills were identified. The incidence rate in 8–9 year-old children with average IQ who had either weak semantic and/or weak episodic memory was 5.9% and in a linked study of 11–12 year-old was 5.8%. These figures suggest a higher prevalence than had been anticipated. In these cases, there was no global pattern of cognitive impairment; instead memory abilities were impaired with intellect remaining intact confirming that the development of memory can dissociate from other cognitive systems and that developmental amnesia is a further identifiable developmental disorder in the school population.

8.2. Dissociation between semantic and episodic memory

The detailed analysis of memory for the case CL revealed that she had impaired semantic memory for both knowledge of facts and semantic knowledge of words. The results were demonstrated on multiple tasks, including general world knowledge and semantic knowledge of words as assessed by receptive vocabulary and conceptual superordinate questions. In contrast to the deficits in semantic memory, there was no impairment in episodic memory for verbal or visual material, whether assessed by recall or recognition. In relation to word knowledge, there was also no impairment in knowledge of word names, the impairments occurred only on certain tasks when semantic knowledge of the word was required. Vocabulary is established but semantic knowledge of that vocabulary is impaired.

In CL’s case semantic memory was impaired but episodic memory and recognition memory was intact. There have been descriptions of children with amnesia with the opposite memory profile, i.e. whose memory has developed with normal semantic knowledge for fact and words, but who demonstrate poor episodic memory (Gadian et al., 2000; Vargha-Khadem et al., 1997, 2002). This double dissociation between the previous cases of amnesia and CL reported here implies independence in semantic memory and episodic memory during development, with neither dependent upon intact skill in the other. This supports models of modularity within memory development, in which the emerging components of semantic memory and episodic memory have the capacity to develop separately from each other.

The performance of CL, whose memory profile differs from previous cases of developmental amnesia, raises issues concerning traditional stage models of development, in which there is only a single route to competence and progress is made only through sequential acquisition of skills at each stage. In relation to such models, impairment must represent arrested development, but these models cannot accommodate different patterns of acquisition where contrasting selective components of memory are impaired whilst others appear intact. The cases of Vargha-Khadem et al. (1997, 2002; Gadian et al., 2000) and the case reported here cannot all be accommodated within a single stage model of memory development and argue for parallel routes within memory acquisition.

8.3. Acquired cases with similar profiles

Impaired semantic memory but relatively intact episodic memory, in terms of anterograde skills has been described in several cases of acquired memory impairment in adults (e.g. Ellis, Young & Critchley, 1989; Goldberg & Bilder, 1986; Kapur, Ellison, Smith, McLellan & Burrows, 1992; Kapur et al., 1994; Kapur et al., 1997). One such case, LT (Kapur et al., 1992), had impaired semantic knowledge about the world in terms of general knowledge and a retrograde amnesia for knowledge of public events, famous scenes, faces and names. Vocabulary was low in relation to other skills and there were naming difficulties. The semantic difficulties therefore extended beyond world knowledge to knowledge of words. In contrast, episodic memory for lists of words was normal. Immediate story recall was less competent than we report for CL but the authors attribute this to the effect of the naming and language difficulties. In common with CL, recognition memory for words was normal and immediate recall of designs was also good. In contrast to CL, recognition memory for faces on the WRMB was moderately impaired. LT also had loss of pre-injury autobiographical
memory though memory for post injury events appeared intact. The authors note that LT had anterior temporal damage but that the hippocampi and limbic diencephalic system appear to be intact. A further case, TJ (Kapur et al., 1994) with bilateral temporal pathology, had impaired semantic memory, including difficulty in establishing new semantic memories but intact episodic memory except for impaired memory for faces and mild weakness in story recall (25th centile performance). Cases of this sort contrast with the significant acquired episodic impairment reported frequently in those with hippocampal lesions such as the classical case, HM (Scoville & Milner, 1957). Nestor, Graham, Bozat, Simons & Hodges (2002) discuss frontotemporal dementia in which there is early sparing of the hippocampus. They distinguish between frontal variant frontotemporal dementia in which semantic memory is preserved but episodic memory is impaired and temporal variant frontotemporal dementia in which semantic knowledge is impaired but episodic memory is intact. Thus a number of studies of acquired amnesia in adults with profiles similar to CL implicate the parahippocampal cortices and the temporal lobes as the substrate for semantic memory.

8.4. Formation of semantic memory

Hintzman (1986) argued that semantic memory is an emergent property of episodic memory. Over a period of time, common elements of repeated episodic memories form a semantic memory. The context of the learning, then becomes redundant. Squire (1992; Squire & Zola, 1998) has argued that through a process of consolidation storage becomes independent of a limbic-diencephalic system. If this were true, then semantic memory problems could exist independent of the functioning of episodic memory, but difficulties in episodic memory would always co-occur with poor semantic memory skills. If semantic memory were thought to emerge from episodic memory by the processes of abstraction and generalisation outlined above, CL could be seen as having normal episodic processes but a failure in this later consolidation or abstraction. The profile of CL for visual learning, in which initial recall is intact but delayed recall is impaired would be compatible with this sole route of entry theory. However, the theory fails to explain why verbal learning and delayed recall is normal for CL yet semantic memory for facts and words is impaired unless a longer timescale for consolidation is envisaged. A further adult acquired case of impaired semantic memory but intact episodic memory was described by Kapur et al. (1997) for whom there was evidence of a longer time-scale to memory loss. In this case, there was loss of semantic memory for news events. In contrast, there was good episodic memory with normal immediate recall and recall after a half hour delay, however, after 40 days there was a dense amnesia for this information. Kapur et al. (1997) argue that the case would be consistent with a multiple stage consolidation process. The case was associated with temporal lobe epilepsy and left temporal lobe pathology, with possible left anterior hippocampal involvement.

Arguing against the cognitive theory of semantic memory deriving from episodic memory is the data from the children identified by Vargha-Khadem et al. (1997, 2002; Gadian et al., 2000) who have developed competency in semantic knowledge whilst having poor episodic memory. A cognitively similar acquired case in an adult, PS, has been described by Verfaellie, Koseff & Alexander (2000). PS had severely impaired episodic learning but relatively preserved semantic learning though this was most evident on tasks with minimal retrieval demands. In both, these child and adult cases (Gadian et al., 2000; Vargha-Khadem et al., 1997; Verfaellie et al., 2000) there is evidence of hippocampal pathology but intact subhippocampal cortices & adjacent cortical areas. Thus, in children and adults with competent semantic memory but impaired episodic memory there is hippocampal pathology but intact adjacent areas. In contrast in the adult cases that parallel CL, with impaired semantic memory but relatively intact episodic memory there may be largely intact hippocampus but parahippocampal and temporal lobe abnormalities.

8.5. Procedural memory

In contrast to the intact episodic memory skills, procedural memory was impaired for CL, with performance becoming more markedly abnormal as trials progressed. This appears to be different from Julia (Temple, 1997) for whom episodic memory was severely impaired but procedural memory appeared to be intact, although the measures of procedural memory in the two studies differ. Maurer (1992) has also demonstrated intact procedural skills in the face of episodic impairment in developmental amnesia and CL appears to form a double dissociation with this case. This implies that all three components of memory—semantic, episodic and procedural, may have distinct developmental pathways which are not mutually dependent and, therefore, may be modelled within a modular system.

8.6. Issues of literacy

CL had impaired reading and spelling. Reading was characterised by significant difficulty in reading irregular and exception words aloud (pint, yacht) and significant confusion in determining the meaning of homophones (sale-sail). There are therefore poor lexico-semantic reading skills. There is impairment in semantic memory and the part of the reading system that depends upon a semantic system is impaired. In contrast, CL was unimpaired in reading non-words. This indicates good phonological reading skills and demonstrates that CL has surface dyslexia. Although there was some impairment of regular word reading, this is often seen in surface dyslexia and may be interpreted as reflecting the variable interpretation of some sounds in English, even in regular words and also the absence of the
advantage in reading of having more than one effective system. The intact reading of non-words however demonstrates that the phonological route has developed normally and the disproportionate difficulty in irregular over regular words reflected in the significant regularity effect in reading confirms this. It is unclear whether cases of amnesia affecting the semantic system in children will always be associated with surface dyslexia. If so, it suggests an interdependency between developing semantic systems. Castles & Coltheart (1996) have demonstrated that children with surface dyslexia need not have visual memory impairments so if a dependency is established it may not be bidirectional. In spelling, phonological skills are again well established but there is impairment in the spelling of both regular and irregular words. There is impairment in the lexico-semantic spelling system. In English, the ambiguities of vowel spelling mean that word specific knowledge is particularly important to differentiate between multiple possible regular spellings and the impairment in spelling regular words as well as irregular words is not therefore incompatible with this interpretation.

8.7. Preservation of arithmetical knowledge

In contrast to the impaired development of the lexico-semantic reading system, semantic knowledge of numbers was intact, at least in relation to the components assessed here. Despite pervasive impairment in semantic knowledge of world facts and knowledge of words, knowledge of numerical facts was unimpaired. This parallels the knowledge of world facts and knowledge of words, assessed here. Despite pervasive impairment in semantic knowledge of numbers and semantic knowledge for the world and words. A relatively intact hippocampus but temporal lobe atrophy was noted in the Butterworth et al. (2002) and Cappelletti et al. (2001) case.

8.8. Residual normality

Despite the semantic memory impairment for CL in domains other than number, the intact episodic skills are at a completely normal level even in comparison to controls all of whom are of average intelligence. The difference in skill is not one of relatively greater deficit in semantic than episodic memory but of significant semantic memory impaired and completely intact episodic memory development. This supports theories of residual normality in developmental disorders (Clahsen & Temple, 2003; Temple & Clahsen, 2002) where the remaining memory skills correspond to components of a normal functional architecture rather than development which is has an atypical trajectory in all domains. There may be multiple routes to competence within these memory systems.

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