

# 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 knowledge 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. © 2003 Elsevier Ltd. All rights reserved.

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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, Szekely, & 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

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disorders may take several different forms and have been explained in relation to selective impairment in the development of components of modular systems underlying face processing and visual perception (Temple, 1997a,b). If modular organisation of the sort argued for in these studies is a fundamental underlying principle in brain development, then one would expect to see focal developmental disorders in all cognitive domains. The current paper concerns a case study of such a disorder of memory.

### 1.2. *Acquired versus developmental amnesias*

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).

### 1.3. *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 known 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 developmental 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.

### 1.4. *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).

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