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Agnosia, alexia and a remarkable form of amnesia in an adolescent boy

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Summary

Childhood cases of global anterograde amnesia, visual agnosia or alexia without agraphia, either alone or in any combination, are extremely rare. Here we report the case of a male adolescent, Neil (a pseudonym), who consequent to a pineal tumour began to exhibit all three disorders in the presence of normal verbal intelligence. The most surprising aspect of Neil's case, however, is his ability to retrieve postmorbid memories through the act of writing without being able to provide any oral account of the content of his written reports. His memory retrieval thus has some of the character of 'automatic writing'. This evidence pointing to Neil's possession of a dissociated form of episodic memory presents a new challenge to our understanding of the organization of memory and of the cerebral systems underlying it.

Key words: agnosia; alexia; amnesia; paediatric; automatic writing

Introduction

The neuropsychology literature on the effects of brain injury in adults contains numerous descriptions of severe, yet isolated, high-level cognitive defects, such as aphasia, amnesia, agnosia, alexia or apraxia. By comparison, the literature on such defects in children is surprisingly limited [aphasia (Landau *et al.*, 1960; Vargha-Khadem *et al.*, 1985); amnesia (Ostergaard, 1987; Wood *et al.*, 1989); agnosia (McConachie, 1976; Young and Ellis, 1989)]. The cognitive impairments that have been reported extensively in children are of a more general type, including mental retardation, autism, learning disability, etc. Even when the reports of impairment in children concern more selective aspects of cognition, the disorder is commonly described as relative rather than absolute, as in dysphasia, dyslexia or dyscalculia. The explanation for this difference between the cognitive effects of brain injury in children and adults is still unclear. One possibility is that during maturation the cerebral foci or 'modules' for specific cognitive functions develop only gradually out of an initially distributed cerebral organization of these functions. Another is that focal brain damage as a result, for example, of stroke, tumour, or even head injury, occurs less frequently in children than in adults. A very different alternative must also be considered, however, namely, that investigation of brain-injured children has simply lagged behind that of adults, the difference between them being one not just of incidence but also of sampling. In our own experience at the Great Ormond Street Hospital for Children, a tertiary referral centre in London, we have now encountered several cases of severe yet isolated high-level cognitive defect in childhood (Patterson *et al.*, 1989; Vargha-Khadem and Isaacs, 1991, 1992; Vargha-Khadem *et al.*, 1991; Incisa della Rocchetta *et al.*, 1992), supporting the possibility that more intensive sampling will yet reveal an incidence of such impairment in children that more closely approximates that already found in adults.

The present report is a description of one such case, an adolescent boy we shall refer to as Neil, who suffered from a metastasizing pineal tumour at the age of 13. The cognitive syndrome that resulted is highly unusual in at least two respects. First, despite retaining normal verbal intelligence, Neil exhibits not just one high-level impairment but three–global anterograde amnesia, visual agnosia and alexia. Secondly, although his amnesia is truly profound in all sensory modalities, he is able to retrieve postmorbid memories through writing without having any awareness, at least to oral report, of the content of his written report. This remarkable phenomenon, which was uncovered accidentally during the course of the investigation, is described below following an

account of his medical history and the subsequent neuropsychological assessment. A brief report of this case has already appeared (Vargha-Khadem and Isaacs, 1990).

Case report

Medical history

Informed parental consent was obtained for the investigation of Neil. Neil was born on January 3, 1974, by a breech delivery at 42 weeks of gestation. His birth weight was 7 lb 6 oz. Neil suffered complications from meconium aspiration and he was in an incubator in a special baby unit for 6 days. Despite the perinatal complications, Neil's early development was normal, and he achieved developmental milestones appropriately. He was always considered a clever child, if somewhat clumsy. He was strongly right-handed. His only boyhood illness of note was a sinus infection at age 12 years, which required surgery.

The first clinical signs of his current disorder appeared in April 1987 when Neil was 13 years old and in his last year in preparatory (junior high) school. These early signs consisted of insatiable thirst despite excessive liquid intake, and frequent urination. He also underwent a gradual change in personality: having been gregarious and outgoing before he had now become quiet and withdrawn. In addition, although he had always done well academically, he failed the Common Entrance Examination. Signs of disorientation and memory loss began to emerge around this time, with Neil often returning home from school missing many of his personal belongings. On one occasion he came home with only one shoe, explaining that he had lost the other while changing into his sports outfit.

In September 1987 Neil transferred to a boarding senior school where he lived for one term. When he visited his family during the mid-term break, his mother noted that he was walking with an abnormal gait and was having difficulty negotiating stairs and obstacles. Also, he was observed examining objects and pictures, including highly familiar ones, unusually closely and for long periods.

When Neil returned home for the Christmas holidays, he brought along his work-books covering the first term; it was noteworthy that they contained nothing written by him during that entire period. The school report expressed concern on the part of his teachers, who had observed several episodes in which Neil had appeared confused, disoriented or forgetful. At the time, however, it was thought that these failures might simply be due to emotional difficulties Neil was having in living away from home and in adjusting to his new school.

During the Christmas holidays, the family travelled to Canada to visit relatives, at which time they also visited Niagara Falls. Neil recalls the outbound transatlantic flight but little else about this trip, except for a most frightening feeling of claustrophobia and dizziness experienced when he was taken in a boat to view the Niagara Falls from beneath them. Neil cannot recall the return flight to England or any event thereafter.

In April 1988, after further deterioration in his fluid regulation, gait and vision, a tumour in the region of the pineal body was suspected, and Neil was referred for medical diagnosis.

Ophthalmological examination revealed bilateral papill-oedema and a visual acuity of 6/36 in each eye. Although his visual fields were normal and his pupils were reactive to accommodation, they were dilated and fixed to light. He also had complete vertical gaze palsy and jerky, horizontal following movements. Other cranial nerves were normal.

His somatosensory and motor status were also normal, except for his gait, which had by now become profoundly ataxic and characterized by a wide stance. The general physical examination was unremarkable except that there was no evidence of secondary sexual development.

A CT and a follow-up MRI scan (Fig. 1A, C) revealed moderately enlarged ventricles indicative of obstructive hydrocephalus caused by an extensive intraventricular tumour. A solid mass lesion in the pineal region of the posterior third ventricle, the presumed primary site, infiltrated the dorsal aspect of the tectum and compressed or infiltrated the medial thalamus. A separate mass, multilocular and predominantly cystic, was present in the anterior third ventricle with extensions into the anterior corpus callosum, cavum septum pellucidum, hypothalamus and suprasella cistern. In addition, there was evidence of extensive oedema involving, posteriorly, the right cerebellum through the right cerebellar peduncle, and, anteriorly, the white matter of the medial halves of both frontal lobes, the forceps minor of the corpus callosum and the subependymal walls of the lateral ventricles.

A stereotactic biopsy was immediately carried out, and this disclosed a pineal germinoma which was verified histologically. The tumour was treated with chemotherapy (carboplatin, vincristine, and bleomycin and cisplatin, over a period of 4 weeks), followed by cranio-spinal irradiation (3000 rads to the whole brain over a period of 3.5 weeks, 1000 rads to the tumour site over 8 days and 3000 rads to the spinal area over 7 weeks). This tumour-treatment regimen was completed in August 1988.

On examination in December 1988, 4 months post-tumour treatment, Neil was found to have a functional disconnection between the hypothalamus and the pituitary body. Growth hormone, thyroxine and hydrocortisone replacement therapy were therefore commenced, and the regime for the control of diabetes insipidus was continued. His delayed secondary sexual development was treated with testosterone, and he was operated on for an undescended testis in April 1989.

Magnetic resonance imaging scans repeated both 1 and 2 years after radiation treatment indicated complete resolution of the tumour but numerous residual abnormalities (Figs 1B, D and 2). These included, in addition to the persisting but decreased hydrocephalus, significant atrophy or thinning of the anterior corpus callosum, the fornices, the grey matter in the region of the medial diencephalon, particularly anteriorly and in the floor of the hypothalamus, the left hippocampal

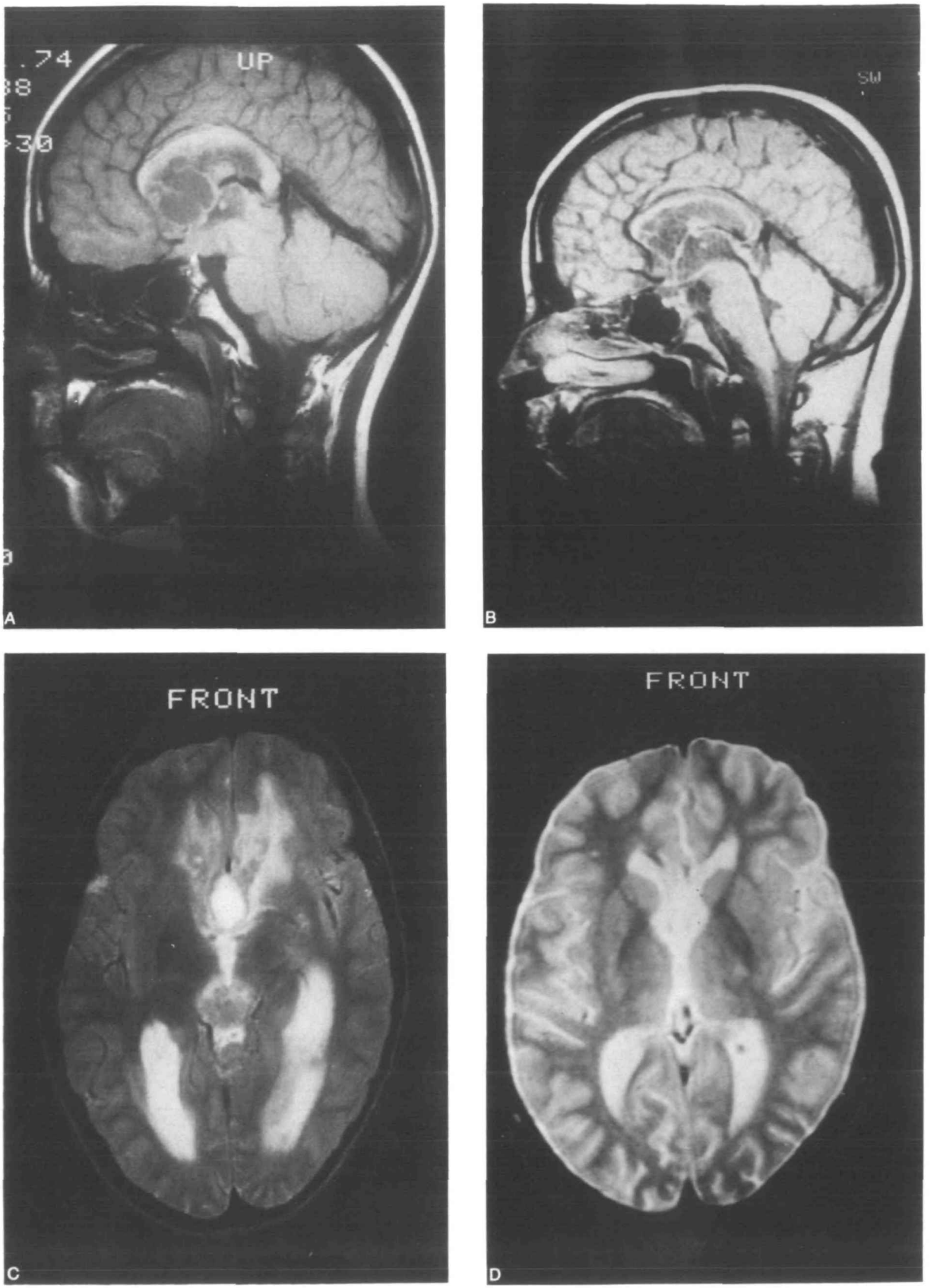

Fig. 1 MRI scans. **A** and **C** are mid-saggital and horizontal sections, respectively, taken at the time of diagnosis (April 1988). **B** and **D** are mid-saggital and horizontal sections, respectively, taken at a different MRI centre about 2 years post-radiation treatment (July 1990). For description of pathology, *see* Case report, Medical history.

Fig. 2 Coronal MRI section, taken about 2 years post-radiation treatment, showing enlarged ventricles, thinning of anterior corpus callosum and atrophy of left hippocampal formation, and, possibly, right inferior parietal lobule. Right hemisphere appears on the left.

formation, the superior colliculi and the right cerebellar hemisphere.

In January 1989 Neil entered a school for the physically disabled. He was registered as legally blind and given a walking stick, which he was successfully trained to use. He was also provided with a closed circuit television monitor that enlarged printed material as an aid in reading, but this proved unsuccessful and was discontinued. Later, he was given access to a computer with an enlarged keyboard and screen, and a start was made in teaching him both touch typing and Grade I Braille, on which he was reported to have made good progress. A dictaphone and tape recorder were provided as aids to memory.

Clinical referral for neuropsychological evaluation

In September 1989, 13 months after radiation treatment, Neil was referred to us for neuropsychological investigation. His neurological status at this time was largely unchanged, except for improved eye movements, including some return of upward gaze. The ataxia, complex nystagmus and extensive visual field loss remained, useful vision being restricted to the nasal hemifield with the left eye and the lower nasal quadrant with the right. As a result he lacked stereopsis (measured by the titmus and TNO tests for stereoscopic vision–*see* Test for Stereoscopic Vision, 1972) and he had little colour vision (Ishihara test).

The clinical mandate in the referral request called for both delineation of his cognitive strengths and weaknesses, so that realistic targets could be set for his academic progress, and the design of a management and rehabilitation programme that could alleviate, if not treat, his complex perceptual and learning deficits. The presenting neuropsychological problems at the time of the referral were (i) marked personality change, (ii) severe visual impairment, including difficulty in reading, and (iii) marked memory impairment. This assessment which was based, in part, on a formal evaluation of his cognitive functions carried out 4 months post-treatment, was made available at referral as part of his clinical documents. To re-evaluate his neuropsychological status, and to attempt to gain an understanding of his complex syndrome, we examined Neil for a total of ~50 h distributed across 18 sessions during the following year.

Personality and communication skills

Neil was found to be a pleasant, polite and cooperative young man with some limited insight into his disabilities. Despite this insight, he was exceptionally placid, rarely demonstrating any emotion or expressing any wishes. At no time in the ensuing year did the examiners see him laugh or smile, become angry or irritated, or display any enthusiam. He seldom initiated a conversation, although he seemed happy to answer when questioned. Being articulate and facile in such verbal exchanges, and so having good conversational skills, Neil's cognitive impairments are not readily noticeable. He is described by his parents as being extremely easy to live with because he rarely makes any requests. Indeed, his mother said she often feels his head is empty, with no thoughts, goals or plans.

Language and verbal fluency

Neil's language function, assessed with selective subtests from the Boston Diagnostic Aphasia Test (Goodglass and Kaplan, 1983), was found to be normal (Table 1). Also, on the Chicago Word Fluency Test (Thurstone and Thurstone, 1962), he generated a normal number of words (39 beginning with S and eight beginning with C, in 9 min), although his responses contained a high proportion of perseverations (13 out of 60).

Intelligence

Wechsler intelligence scale for children-Revised (WISC-R). Neil's IQ was first measured 4 months after radiation treatment (during the assessment prior to his referral) and again 18 months after treatment (Table 2). Verbal and performance IQs were highly discrepant in both testing

Table 1 *Boston Diagnostic Aphasia Test*

Subtest	Raw scores
Body part identification/right-left discrimination	18/20
Execution of commands	15/15
Production of automatized sequences	8/8
Repetition of words/articulation	10/10
Repetition of phrases	
High probability	8/8
Low probability	7/8
Fluency (generation of animal names)	26/19*

*Standard maximum

Table 2 *Intelligence Quotient (WISC-R)*

	1988	1990
Verbal IQ	**111**	**109**
Information	13	12
Similarities	12	9
Arithmetic	9	10
Vocabulary	12	14
Comprehension	13	13
Digit Span	—	5
Performance IQ	**71**	**55**
Picture Completion	5	1
Picture Arrangement	5	—
Block Design	8	10
Object Assembly	4	1
Coding	—	1
Mazes	—	1

sessions. Verbal IQ was just above the average range in 1988 and declined only two points by 1990. In contrast, performance IQ was already at a borderline deficient level in 1988 and had deteriorated still further by 1990, falling well within the mentally deficient range. Yet he obtained scaled scores of eight out of 19 and 10 out of 19 (i.e. average performance for his age) on the Block Design subtest of the performance scale, suggesting that his visuospatial perception was relatively intact. By implication, his extremely low overall score on the performance scale is unlikely to have been due either to his primary visual defect or to a markedly deficient non-verbal intelligence.

Tower of London. (Shallice, 1982). Neil was given two simplified versions of this puzzle (three objects graded in size with three locations for stacking them, and four objects with four locations) before receiving the standard version (four objects, three locations). In each case, after just one or two trials, he achieved problem solution in the minimum number of moves (i.e. seven, nine and 17 moves, respectively). Besides providing another indication of normal non-verbal intelligence, the result demonstrates that he has sufficient visuospatial perception to process, at the least, both object size and location.

Memory

Autobiographical memory, retrograde. A list of significant persons, places and events spanning Neil's life from about the age of 4 to 12.5 years was compiled in consultation with his parents. Queried about each of these items, Neil exhibited virtually perfect memory.

Autobiographical memory, anterograde. A similar list of significant items was compiled spanning Neil's years from 13 to 16. Questioned about names and episodes within this period, he failed completely. In an attempt to document the frequency of Neil's everyday memory problems, his mother was asked to fill out two different questionnaires [An Inventory of Memory Experiences (Herrmann and Neisser, 1978) and the Memory Questionnaire (Sunderland *et al.*, 1983)]. Her responses indicated that Neil nearly always forgets everyday items, including appointments, errands, planned activities, object locations, taking personal belongings, directions to new places, names, telephone numbers, shopping lists, conversations and participants, television programmes, songs, etc. Neil himself is fully aware of these everyday memory failures, for when the questions were read to him he answered in essentially the same way his mother had. When asked to describe the types of activities his memory failure interfered with most seriously, he replied, 'School. Remembering what I have learnt. If I ask the teacher a question, by the time she has answered me, I have forgotten what I have asked.' Both Neil's and his mother's responses to items pertaining to his premorbid memories, however, indicated that these were quite intact.

Temporal orientation. Neil's temporal orientation had also become seriously impaired. On initial testing at 4 months post-treatment, he was described as being well oriented in time and place. On retesting 14 months later, he knew the year, but not the month, date, day or time.

Memory quotient. In the assessment 4 months post-treatment, Neil had been administered the Adult Memory and Information Processing Battery (Coughlan and Hollows, 1985), the norms for which begin at the age of 18 years. On the mnemonic measures of this test, which are similar to the ones contained in the Wechsler Memory Scale (WMS), Neil's scores fell within the impaired range. These measures included list learning (28 out of 75 words), figure recall (immediate, 49%; delayed, 43%), design learning (29 out of 45 and one out of nine elements) and speed of information processing. On story memory, he achieved a just-below average score (30 out of 56 elements) on immediate recall, but then fell once more within the impaired range (20 out of 56 elements) on delayed recall. Only on the non-mnemonic measures of verbal fluency, a maze test, and cube drawing did he perform within the normal range.

Neil's Memory Quotient on the Wechsler Scale (Wechsler, 1945), measured when he was 14 months post-radiation treatment, was 59 (Table 3), indicative of a further, serious

Table 3 *Memory quotient (Wechsler Memory Scale—Form ii)*

Memory quotient	**59**
(1) Personal and current information	83%
(2) Orientation (temporal and spatial)	60%
(3) Mental control	56%
(4) Immediate recall of prose passages	13%
(5) Ninety minute delayed recall of prose passages	0
(6) Digit span	
Forwards	3
Backwards	2
(7) Visual reproduction	44%
(8) Forty minute delayed recall of visual reproduction	0
(9) Paired associate learning	16%
Related pairs	3/10
Unrelated pairs	0/10
(10) Ninety minute delayed recall of paired associates	0

decline in his memory ability. Because this scale consists primarily of verbal items, the resulting memory quotient usually correlates closely with verbal IQ. In Neil's case, however, there was a discrepancy of 50 points between the two, indicating a severe verbal memory deficit in the presence of normal verbal intelligence. His memory for visual information was, of course, also profoundly impaired (*see* Table 3, Visual reproduction), but in this case it was unclear how much of the impairment was due to memory failure and how much to his defective vision. Additional tests of memory were therefore administered in the auditory and tactual modes.

Auditory memory. On the Rey Auditory Verbal Learning test (Rey, 1964) Neil recalled a total of ~20% (17 out of 75) of the words from list A (15-word list, repeated for five trials) and one word from list B (15-word list, one trial). These scores fall below the tenth percentile for 5-year-olds. On 30 min delayed recall he remembered none of the words, although he did remember having been read some word lists. He performed somewhat better on the 30 min delayed recognition test, correctly identifying as words he had just heard 80% (12 out of 15) of the ones on list A and 33% (five out of 15) of those on list B; of the distractors, he incorrectly identified as words-just-heard only 17% (four out of 30).

Ninety percent of the few words that Neil had recalled on the initial phase of the Rey test were from the last four words of each of the two lists, and in each case he reported the final word first. To determine the reliability of this marked recency effect, six lists of 10 high-frequency words were presented once each, with essentially the same result: eight of the nine words that Neil recalled were from the last three words in each list, and again in each case he reported the final word first.

Single or double consonants were presented by the Brown–Peterson method (Brown, 1958), i.e. with delays that were either unfilled or filled with counting backwards. Neil correctly recalled single and double consonants after unfilled delays of up to 9 s, the longest tested, but he could not recall even single consonants after filled delays as short as 3 s.

Pairs of unfamiliar melodies, each consisting of two 9 s segments separated by 2–3 s, were presented through earphones binaurally, and Neil was asked whether or not the second segment matched the first. He scored at the level of chance (25 out of 50). This failure was not due to an inability to recognize non-verbal auditory material, since he easily identified familiar environmental sounds (e.g. horse neighing, audience clapping), scoring ~90% correct (16 out of 18).

Tactile memory. Five objects that could not be easily named (Lego blocks) were presented for tactile exploration for 5 s each, after which these five objects were intermixed with five others for recognition. On this test Neil scored at chance (five out of 10) with the left hand, and then, with the same objects but a different set of targets, he scored perfectly (10 out of 10) with the right.

Ten pairs of abstract wire shapes were presented, one pair at a time, the first shape in each pair for tactile exploration for ~10 s, followed immediately by the second of each pair for same/different judgement. Six such sets, with new shapes for each, were presented to one hand or the other in counterbalanced order. The three scores for the right hand were 80, 90 and 90%, whereas those for the left were 40, 0 and 50%. As with his failure on the music recognition test, Neil's failure in tactile shape recognition with the left hand was not due to any perceptual impairment. He correctly identified within 1–2 s each of 16 common objects (comb, key, ball, spoon, coin, pencil, etc.) placed in this hand, just as he could with his right hand. Imposition of even a short delay, however, resulted in impaired recognition of the wire shapes with either hand. Thus, when a 10 s interval separated exploration and choice, Neil scored 70% on one set with the right hand and 50% on another with the left. He also performed near chance on a continuous recognition test in which wire-figure targets (the repeated items) were separated by one, five or 10 distractors, each adding a delay of ~10 s. Different sets of figures were again given to the two hands. On this test Neil performed no better with one distractor than with 10, and no better with the right hand than with the left, scoring 60% (43 out of 72) with each. However, he responded 'familiar' more often to both target and distractor items placed in the right hand (44%) than he did to items placed in the left (26%).

Visual identification

As noted earlier, it was unclear how much of Neil's impairment on the Visual Reproduction Subtest of the Wechsler Memory Scale, as well as his everyday visual recognition and recall failures, was attributable to his memory loss and how much to his defective vision. His ability to identify visual stimuli was extremely poor. For example, he was unable to name familiar objects located in an unfamiliar setting, including placement on a table directly in front of him. Tested visually in this way with the common objects

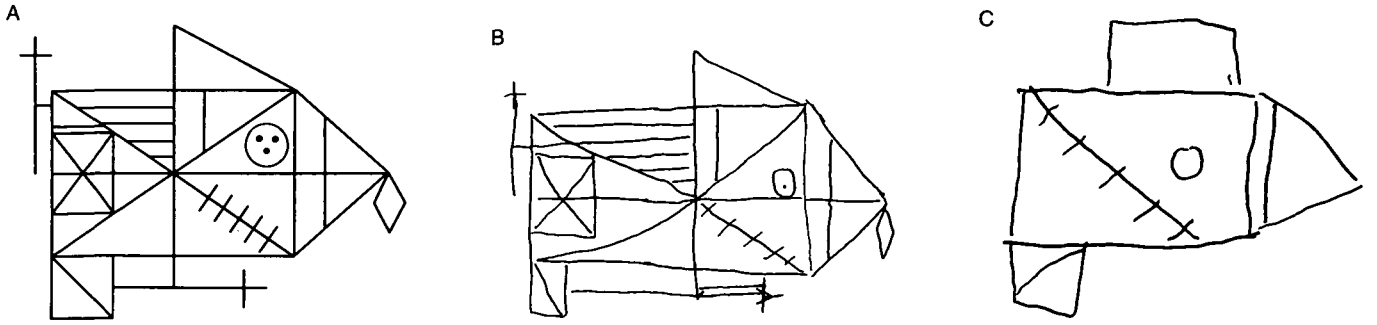

Fig. 3 (**A**) Rey–Osterreith complex figure. (**B**) Neil's copy. (**C**) His reproduction from memory, 40 min later.

that he could readily identify tactilely (*see above*), he named none of them, even when clued about their use. He succeeded only when familiar objects were in their customary places, as with his personal belongings and furnishings in his home. Similarly, he could only guess at the identity of famous faces. Additional quantitative evidence of his impairment in visual identification were his scores of 20% (two out of 10) on the first 10 line-drawings of the Boston Naming Test (Goodglass and Kaplan, 1983), and 39% (11 out of 28) in identifying common household objects from large achromatic photographs (mean latency of 37 s to first response). In both cases, when the objects were orally defined instead, he named them quickly and accurately. Neil's somewhat better performance with pictured than with real objects suggests that light and depth cues may actually have interfered with his perception. His identification of the photographs was not aided by semantic cues, but was by phonetic cues, though even then he often denied that the pictured objects looked like the ones he had just correctly named. Not only was he poor at naming pictured objects and famous persons, he was also unable to identify emotional expressions portrayed by unfamiliar faces, scoring at chance (24 out of 48) on Young and Flude's (1989) test of this ability. The one exception to this impairment in picture identification was his score of 80% (eight out of 10) in naming familiar places, relatives and friends in photographs taken from an old family album.

Visual perception

In an attempt to determine the basis of Neil's visual identification impairment, a series of match-to-sample tests was administered. On two different face-matching tasks (Benton *et al.*, 1983; Young and Flude, 1989), Neil obtained only chance scores (11 out of 27 and 25 out of 48, respectively). On two different spatial-matching tasks, however, his performance improved substantially. Thus, on a test requiring same/different judgements of photographs of three-dimensional abstract shapes (tinkertoys; Layman and Greene, 1988), Neil performed perfectly (five out of five) when the two shapes were in the same orientation, dropping to chance (four out of eight) only when their orientations differed. And again, on judgement of line orientation (Benton *et al.*, 1983), he obtained the surprisingly high score of 90% (54 out of 60), and each of his errors deviated from the target by only one segment (18°). Another example of Neil's visuospatial ability is provided by his performance on the Rey–Osterrieth Complex Figure Test (Osterrieth, 1944), which was shown in original size and contrast (Fig. 3A). Figure 3B illustrates Neil's copy, which was scored 29 out of 36 (the norm for 12-year-olds; Kolb and Whishaw, 1990). In view of his severe visual limitation, the accuracy of his drawing is noteworthy. But, as expected, his reproduction of the drawing 40 min later (Fig. 3C) was markedly degraded (score of 6.5 out of 36, below the norm for 6-year-olds).

Neil's *immediate* memory for visuospatial material was considerably better. This assessment is based on his performance on a modified test of the Corsi Blocks, on which he showed a forward span of six (the norm for 15-year-olds; Isaacs and Vargha-Khadem, 1989).

Drawing

Remarkably, despite his profound deficits in processing, identifying and recognizing visual objects, Neil demonstrates considerable skill in drawing. He produces precise and even intricate renditions of imagined objects and scenes. A few of these taken from his sizeable collection are reproduced in Fig. 4. Yet, only seconds after completing a drawing, he is unable to identify its contents or even that it is his own.

Reading

Neil's reading ability was formally assessed three times, in December 1988, October 1989 and February 1990. On the first occasion, 4 months post-radiation treatment and at the chronological age of 14 years 11 months, his reading age (Graded Word Reading Test; Schonell, 1974) was 11 years 8 months. On a test of prose reading (Neale Analysis of Reading Ability, Form A, Everest; Neale, 1958) given in this same session, his reading age was 11 years 10 months (130 out of 139 words read correctly). As expected from his profound memory disorder, however, he was later unable to answer any questions about this eight-sentence story.

Fourteen months post-treatment, at the chronological age of 15 years 9 months, Neil's reading age had declined to 7 years 3 months (Graded Word Reading Test; Schonell, 1974).

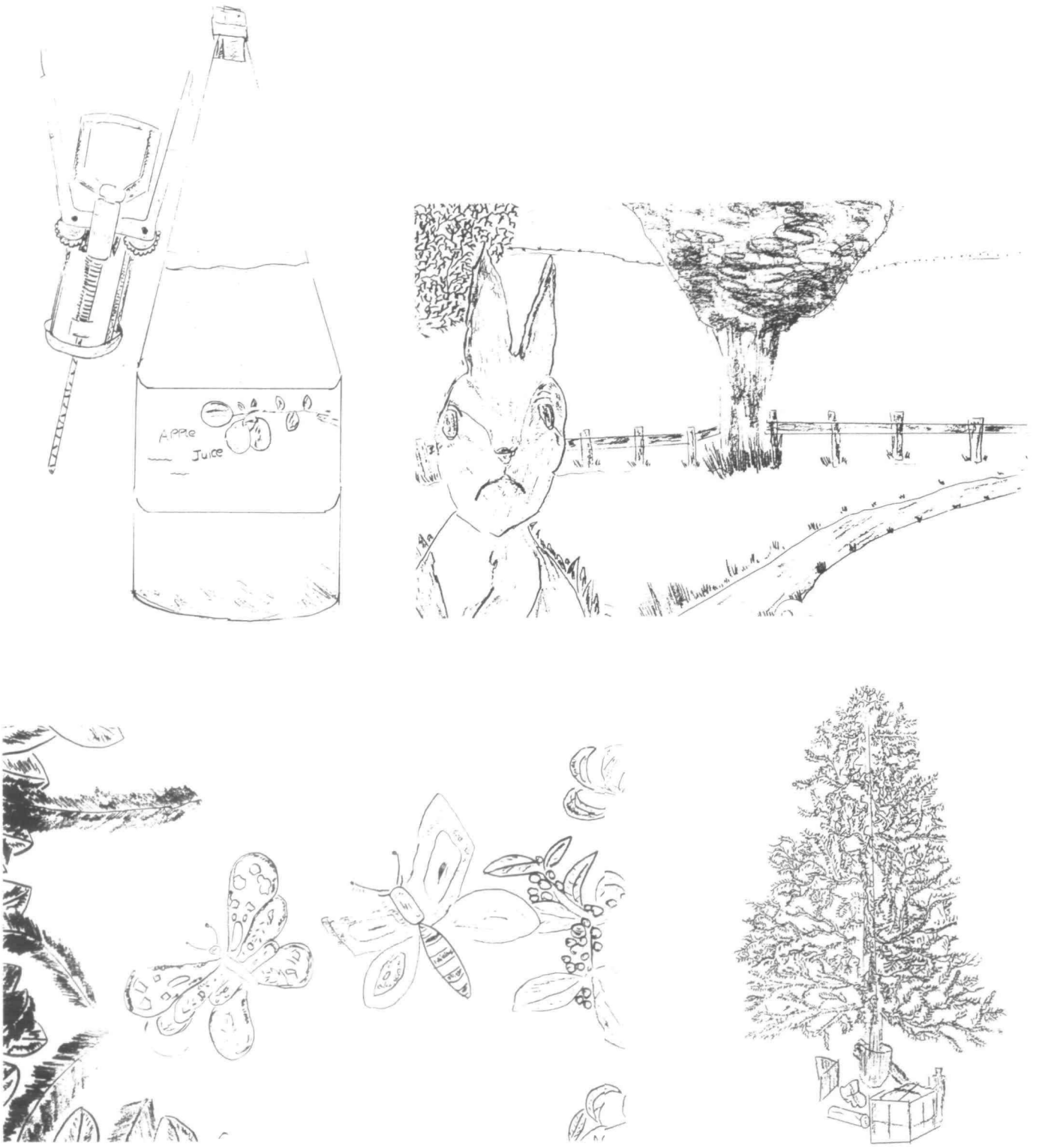

Fig. 4 A sampling of Neil's drawings.

Still later, at 18 months post-treatment, Neil could no longer identify even single numerals or letters, or match upper-case letters with lower-case, although he could discriminate letters from each other and also both trace and copy them. This dramatic deterioration in reading ability over a 14 month interval was not the result of a progressive loss in primary vision, for his acuity remained relatively constant during this period. Rather, like his disorder in object and face perception, his reading disorder appeared to reflect an agnosia, in this case for words and letters.

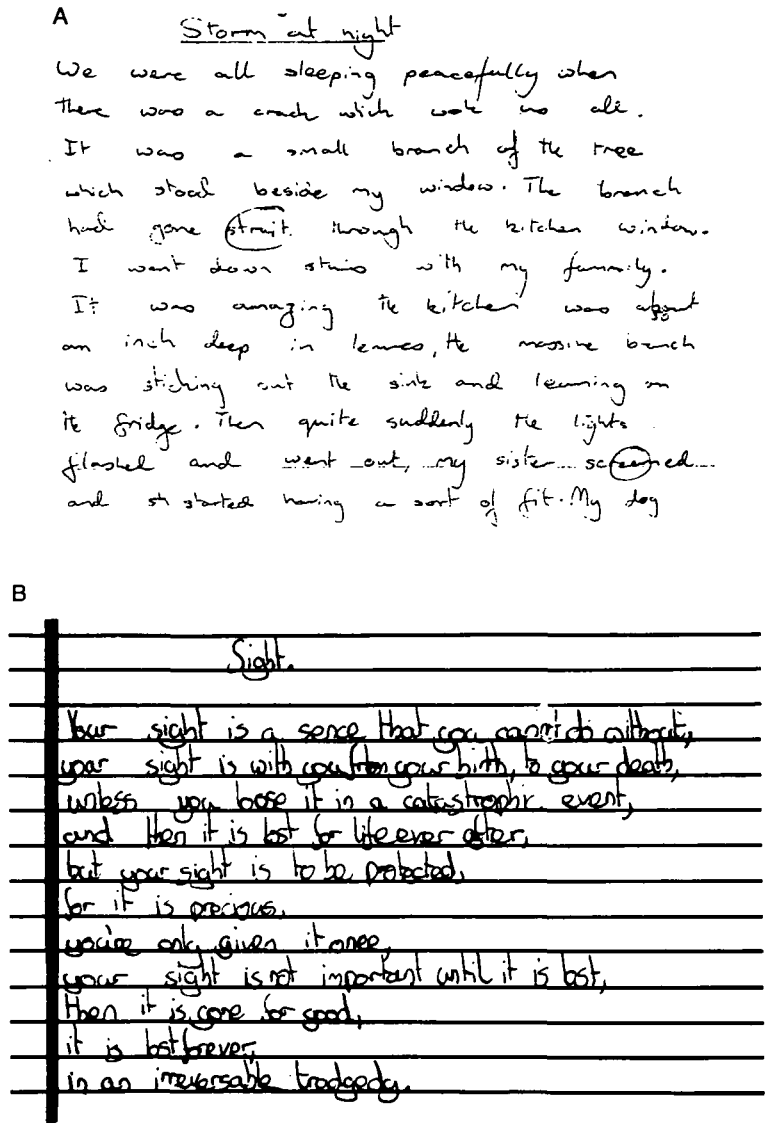

Fig. 5 (A) A sample of Neil's premorbid script. (B) A sample of his postmorbid script.

Writing

Script. Like his verbal IQ and drawing skills, and unlike his reading ability, Neil's writing ability is remarkably intact. His script, which he produces slowly and with effort, consists mainly of lower-case printing (which is, in general, correctly capitalized), but it also includes occasional words and letter-strings in cursive writing. This is the reverse of his premorbid script in which cursive writing predominated. Samples of his earlier and current script are illustrated in Fig. 5A, B, respectively.

Spelling. On a written test of spelling (Single Word Spelling; Schonell, 1942) given when he was 15 years 9 months, his test age was 13 years 6 months, suggesting preservation of the vocabulary he had acquired prior to his tumour. He also did well on two other tests of writing to dictation, scoring perfectly (12 out of 12) when asked to spell both monosyllabic non-words phonetically (e.g. 'roop' and 'dack') and monosyllabic words with irregular spelling (e.g. 'know' and 'gnome'). He did almost as well (14 out of 15) when asked to spell polysyllabic low-frequency words (such as 'explicate' and 'epithet').

Composition. Neil's writing skills extend far beyond the ability to write single words. Just as he produces complicated drawings that he fails to recognize moments later, he is able to produce multi-page essays as well as blank verse, which he can neither read nor remember seconds after completing them. Yet these compositions reflect a smooth flow of well organized ideas and are not repetitious. A particularly poignant piece is duplicated in Fig. 5B. The issue of how he is able to draw multi-object scenes and compose extended passages in the face of his severe memory loss is considered below.

Compensatory strategies involving kinaesthesis

In February 1990, about half-way through the year of testing on which this report is based, it was discovered that Neil could make use of his drawing and writing abilities to compensate in part for his selective cognitive impairments. The first such indication occurred shortly after he realized he could no longer recognize even single letters (*see above*). On the day following that reading test, he attempted to decipher a headline in a newspaper his mother was reading. Frustrated by his failure, he spontaneously began to copy the letters on paper. Realizing he could recognize each letter after copying it, he proceeded to read the headline in this way, letter by letter, word after word (Fig. 6A). Pleased by this success, he immediately went on to copy the outline of a helicopter that appeared below the headline, and identified this too after he had finished drawing the propellers (Fig. 6B). Gradually over the next few days and weeks, copying became Neil's standard strategy both for reading text and for identifying pictured objects, although it did not help him with cursive script or, of course, faces. If, instead of copying, he was asked simply to use his index finger to trace words letter by letter or to trace single objects, he was again able to identify them. He succeeded almost as well if his hand was moved passively around the outline of a letter or a simple object. On one occasion he was asked to trace single letters with eye movements alone, and again he identified them correctly. It was evident that Neil was substituting kinaesthetic and proprioceptive information for visual input, although he himself had no idea how his newly discovered strategy worked.

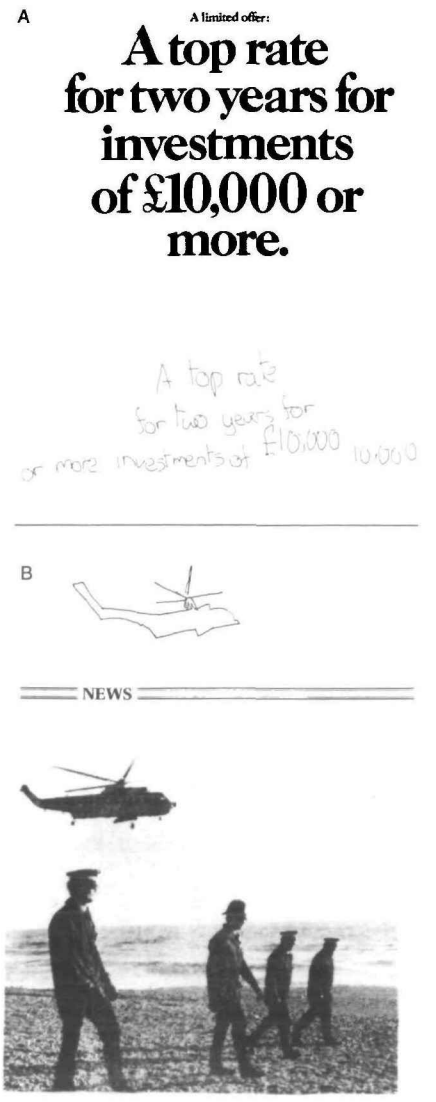

Fig. 6 (**A**) Neil's copy (*below*), enabling him to 'read' the newspaper headline (*above*). (**B**) His outline copy (*above*), enabling him to recognize the helicopter in the newspaper photograph (*below*).

Retrieval of memories through writing

Results of memory testing had demonstrated unambiguously that Neil was suffering from a profound global anterograde amnesia. Yet this conclusion seemed inconsistent with Neil's school reports, which clearly indicated that he was learning and retaining new information, particularly in mathematics and English. In fact, he was completing the standard curriculum at his new school preparatory to writing national examinations in these subjects, as well as in geography and art. As indicated earlier, to assist Neil in his studies, the school had provided him with a tape recorder as a memory aid. Typically, he would listen to taped lectures and write out the answers to questions posed by his teachers. Based on these written responses, his teachers rated his progress in all of his studies as satisfactory.

With the aim of resolving the contradiction between these school reports and the neuropsychological findings, an attempt was made to determine precisely how much new information Neil had actually acquired in his English course. He had been listening most recently to the taped version of the book *Cider With Rosie* by Laurie Lee, and so he was asked specific questions about the book, including the title, none of which he was able to answer. Pressed further he became progressively more frustrated, to a point where he asked to end the session. The examiner agreed pending a final request. Because his grades were known to be based largely if not exclusively on his written responses, he was asked if he could write down the answers, starting with anything at all that he could remember about the book. Initially he did nothing, but at further urging he printed out some words, and, relieved at having finally responded, handed the page to the examiner asking, 'What have I written?' He had printed the words 'Bloodshot Geranium windows Cider with Rosie Dranium smell of damp peppar and mushroom-growth'. The examiner, being familiar with the book, recognized the phrases as ones that came directly from the text. When his words were read back to him, his reaction was one of neither surprise nor pleasure, but rather matter of fact.

Puzzled but encouraged by Neil's ability to retrieve information in this way, the examiner immediately turned to

significant events and persons in Neil's postmorbid experience. Asked whether he could record anything about the events surrounding his hospitalization, he wrote, 'A man had Gangreen', alluding to the fact that, on his hospital admission in April 1988, a man seated next to the driver of the ambulance that brought him to the hospital had a gangrenous foot. Asked for the names of the physicians and hospital staff that cared for him, he correctly wrote the names of several of them. He also wrote the names of several of the children who had been in his hospital ward.

The surprising finding that Neil could retrieve memories in response to questions through writing was discussed with Neil's parents, who up to that time had been frustrated by his inability to recount any event or activity that occurred during his daily life. Later that same evening, his mother urged him to write down the names of children and staff in his new school. He proceeded to produce a long list of these names, all of which were later verified. The next afternoon, in answer to his mother's request to write about what had happened at school that day, he wrote, 'Mum I saw tulips on the way home'. This was the first time in over 2 years that his mother had gained any information by directly questioning Neil about his experiences when he was away from her.

From then on, Neil carried a notebook in which he would enter the answers to questions his parents asked him. All of these entries had to be read back to him, and he remembered none of them afterward. The same was true for the rare instances in which he would write a question, request or reminder of his own. For example, after the test session in which he had listened to music segments through the earphones of a Sony Walkman, he asked for the name of the apparatus and wrote in his notebook that he wished to purchase one. The following day he had to be reminded twice of what he had written before the shopping trip was arranged. On another occasion, in answer to what he had done in school that day, he wrote, 'Today at school I watched the film my left foot [i.e. My Left Foot]. I also had geography. I saw Mr. (name of the teacher) and he was very pleased. Miss (name of the teacher) is going to give us a note about activities in the holidays.' After he wrote this, his mother asked him if had enjoyed the film, to which he replied, 'What film?'

Once, when he was asked to write down the items that his mother had just purchased for him, he wrote, 'She went to buy . . . ' finishing with a tight scribble, after which he passed the sheet to the examiner asking, as he usually did, 'What did I write?' The examiner explained he had not yet answered and handed the sheet back, whereupon Neil wrote, 'some . . .' followed by another tight scribble. He then passed the sheet back and asked 'Did anything come?' Told no, he volunteered 'Let me try again' and then wrote, 'it was a . . .', after which he passed the sheet back a third time. Told once more that he had not yet answered, he abandoned the effort and was not pressed further. That Neil is unable to report orally not only what he has written but even what he is currently writing was confirmed repeatedly.

In writing answers to questions, Neil retrieves information not only about persons and events but also about places. Thus, 'I first discovered that I could use my hand at The Wolfson Center. With my my Mum and Dr. Cadem [Khadem]. I was sitting at the table copying out of a newspaper.' Later, in answer to a repeat of the same question, he wrote, 'I was sitting at the table in front of a serving hatch in the wall. I copied large print from the newspaper. I was in the dining and the sitting room. We were looking onto an Astroturf football pitch.' Again, in a later session, he wrote, 'We just went upstairs and we went into a room with a blind. We did my visual fields and letter seeing. I afterward came down in the lift with Dr. Issacs [Isaacs].' In fact, a different examiner had accompanied him in the lift, but this was his only factual error in these two accounts.

On one occasion, Neil was asked to delay writing until he had been presented with two or three unrelated questions in succession. To some of these questions he was asked to write with his left hand. Out of 20 questions presented in this way, he made only three errors of omission, performing equally well with either hand. In another session, he wrote the correct answer when an 'it' in the current question referred to the main item in the immediately preceding one.

Neil's written responses sometimes contained memories of scenes and objects thought to be beyond his perceptual capability. He was therefore asked to identify in writing six objects and the line-drawings of four others, none of which he could name orally. He correctly wrote the names for a video cassette and a [sea]shell, and, although he could not name it, he correctly described an automatic pencil as green and transparent. He failed to identify an ashtray, doll's comb and pencil case, or any of the four line-drawings, although he succeeded on the latter when allowed, as before, to trace their outlines (*see* Compensatory strategy involving kinaesthetics). It thus appears that writing provides Neil with access to some colour, object and scene vision that he cannot report orally, although the gain is rather modest.

The contrast between his inability to recount recent experiences orally and his ability to do so in writing is well illustrated by the following episode. One afternoon the examiners took him on a 2 h outing, first to a museum, then to a restaurant in a shopping area, and finally for a drive along the shore followed by a walk along it. Neil knew all of these sites from early childhood and directed the examiners to them. On the return trip, when asked to describe whatever he could about the outing, he could recount nothing. On arrival home, he was requested to write about what had just taken place, and he wrote the following, 'We went to the museum [proper name spelt correctly], and we had some pizza. Then we came back, we went onto the Beach and we looked at the sea. Then we came home.' Finally, he was asked to say whether or not certain events had occurred that afternoon, including all those contained in his written account as well as some he had failed to include. He answered, 'Yes', but with hesitation, only to whether he had visited the museum, explaining, 'I just have a slight feeling inside me'.

His memory retrieval through writing was tested quantitatively with forward digit span. When tested orally twice before (*see* Intelligence and Memory) he had obtained spans of five (WISC-R) and three (WMS), respectively. On the occasion in question, he obtained a span of four orally, but seven in writing, the latter being the norm for 15-year-olds tested orally (Isaacs and Vargha-Khadem, 1989). This is the only instance in which he had ever achieved a digit span above five.

Although Neil's memory as assessed through his writing is remarkable, indicating that he has stored information that he is unaware of even while he is retrieving it, the capacity of this memory store appears to be quite limited. During nine different sessions between March and August 1990, Neil was requested on 15 different occasions spread over 8 days to write an account of his recent experiences, as well as to write the answers to over 90 questions pertaining to them (*see* Appendix). None of the written accounts contain references to more than five events each, and none include very much descriptive detail. Also, as indicated earlier, neither these accounts nor his written answers to the specific questions were entirely error free.

Another indication of the limitation in this dissociated memory store was his performance on the free recall of a list of 10 unrelated words. When tested orally, he produced only the final word in the list, just as he had before (*see* Auditory memory). When tested on a second list and asked to respond in writing, he produced a list of seven words, but only two were correct. Of the five erroneous words, one was an intrusion from the preceding list, one ('time') was possibly a phonetic error or composite for two other words ('tie', 'crime') in the current list, and one was the perseveration of a correct word from his written list; no likely source could be identified for the two remaining errors of commission.

Although the capacity of his memory store accessible through writing is clearly limited, it bears repeating that on no occasion could Neil offer an oral account or oral answer, including an unambiguous 'yes' or 'no', to any question pertaining to his postmorbid experiences.

A very different illustration of Neil's ability to retrieve memories motorically but not orally was his successful delayed recall of a series of hand signals (Kolb and Milner, 1981). Neil was asked to lie face up on a sofa and, while viewing the examiner who stood behind him, copy with his left arm and hand a left arm-and-hand sequence that the examiner demonstrated. A series of five movements was built up one at a time, until Neil performed the entire sequence correctly twice in a row at the examiner's signal. He was then distracted with other activities for 10 min, after which he was requested to lie down on the sofa once more. At the previously taught signal, Neil accurately produced the entire movement sequence, at which point he quickly rose from the couch and asked, with a rare display of animation and surprise, 'Where did that come from?' When asked to write about what had just happened, he printed, 'You showed me a sequence with your arms. left. on the settee. The movements are simular to semafore.'

The procedure just described was the last one on which Neil was tested. Further investigation was precluded by a decision of Neil's parents that the year-long evaluation had served its purpose and that additional testing would not further Neil's interests. This decision was based on the parents' assessment that, whereas part of the clinical mandate had been fulfilled, namely, delineation of Neil's cognitive strengths and weaknesses, there was little hope of accomplishing the remainder, which called for the design of a program of rehabilitation. Since Neil's cognitive problems appeared to them now to be both chronic and intractable, they felt that subjecting Neil to further investigation would not be justified.

Discussion

Before Neil's unique pattern of impaired and spared cognitive functions can be considered in detail, we must first address the question of the authenticity of his remarkable amnesic syndrome. This issue needs to be confronted at the outset because Neil's ability to write down memories that otherwise seem totally inaccessible to him, even during and after the writing, is both unprecedented in the literature on anterograde amnesia and unexplainable by current conceptions regarding the neurobiology of memory. Neither of these circumstances alone would necessarily be cause for scepticism about the genuineness of his disorder. Together, however, they provide a powerful reason for considering the possibility that some of Neil's cognitive symptoms are either feigned or, alternatively, functional (i.e. psychogenic) rather than being the direct consequence of his neuropathology.

The feigning of chronic anterograde amnesia has been encountered in legal cases, most often in connection with claims for compensation or benefits following accidental head injury (e.g. Guthkelch, 1980; Pankratz, 1983). That particular motive is of course absent in Neil's case, but another is quite possible, namely, an emotional need to attract the attention or concern of family, friends and teachers. Simulation driven by such a motive might seem to offer a simple explanation of Neil's puzzling syndrome, but that solution faces serious problems of its own. Neil's apparent hypoemotionality, readily interpretable as a sign of cerebral damage, seems to belie any strong emotional need for sympathy or attention, particularly since, as described below, gaining and keeping these emotional supports would demand intense and constant effort. Moreover, simulation would have exacted a high emotional cost. Because his day-to-day memory problems are those of someone who is totally incapacitated, he is forced to lead a life that is severely restricted and constantly supervised, and hence one that to any normal adolescent would surely be frustratingly confining and distressingly repetitious and boring. Yet, from the start of his personality change, Neil became increasingly compliant and his demeanor increasingly placid.

Another difficulty with the notion that Neil feigns his amnesia is his unremarkable intelligence. On both IQ

examinations, carried out 14 months apart, Neil scored near or at an average level on the verbal scale and near or at a mentally deficient level on the performance scale. So large a discrepancy between verbal and performance IQ is rarely encountered except in individuals with brain damage. Moreover, Neil's overall intellectual level seems incommensurate with a perfectly executed strategy of chronic deception. Neil's test performance, assessed during 18 separate sessions spanning a period of 12 months, was extremely consistent. For example, in addition to the reliability in the pattern of his IQ scores, there is the sharp and consistent distinction between his premorbid and postmorbid memories, the former well preserved, the latter essentially absent, to oral recall. Confirmation of a dense anterograde amnesia was also obtained consistently on formal testing. In addition, his scores on digit span, tested on three widely separated occasions, were three, five and four, in that order. Similarly, his immediate tactile recognition with left and right hands, also tested on three different occasions, always showed the same asymmetry of nearly chance performance with the left hand combined with normal performance with the right. Again, in word-list learning, he exhibited a pronounced recency effect on each of many tests, one separated from the others by several months. The ability to plan such a complicated strategy of simulating selective impairment, and then to execute it so consistently throughout the year, would seem to require superior or even exceptional intelligence and memory, not just the average memory, at most, that would be expected in a normal adolescent with but an average verbal IQ.

The notion that Neil is feigning a global memory loss faces yet another hurdle. Only after he had exhibited deteriorating memory over several years, from sporadic forgetting at the beginning of his illness in early 1987 to the full-blown amnesia that characterized his behaviour in early 1990, was the accidental discovery made that Neil could retrieve some of his postmorbid experiences through writing. The motive for suddenly displaying so anomalous an ability would have had to have been subtle indeed, namely, to surprise the examiners. Yet, if that had been his motive, at no time did he evince any pleasure at his success. Moreover, the emotional and intellectual demands of successfully simulating the continued amnesia would now have greatly increased.

Unfortunately, short of gaining an admission from a suspected simulator, deception cannot be proved; i.e. no method has yet been devised that distinguishes accurately between feigned and genuine amnesia (for a comprehensive review, *see* Schacter, 1986). However, one behavioural feature that sometimes discriminates between the two is the tendency of simulators to overplay their role, apparently because of a widely held belief that true amnesics forget more than they actually do. Consequently, in forced-choice recognition tests, where genuine amnesics often obtain scores above chance, simulators may perform only at the chance level or even below (Brandt *et al.*, 1985; Lezak, 1983). By this admittedly imperfect criterion, Neil would probably not be judged a simulator, inasmuch as his scores on the auditory and tactile recognition tests often indicated a significant amount of retention of the sensory information.

Setting aside, for the moment, the possibility of feigned amnesia, which seems to raise many new problems for the one it solves, we turn to the possibility that Neil suffers instead from a chronic form of genuine but functional amnesia. Two different chronic, pathological forms have been described in the literature: functional retrograde amnesia, involving a loss of personal identity and autobiographical memory, and multiple personality disorder, in which the memories of one personality are unavailable to the other(s). (For a comprehensive review of functional amnesias, *see* Schacter and Kihlstrom, 1989.) Since neither of these chronic forms of functional amnesia apply in Neil's case, a totally new form would have to be posited which has the particular amnesic features he exhibits. These would then include, among others, a dense and global anterograde memory loss in the presence of preserved retrograde memory, combined with the mysterious ability to recover some postmorbid memories through writing. In this respect, nothing is gained by such a proposal, since it simply trades the original mystery of a dissociated form of amnesia for an identical mystery differing only in its initial cause.

Of the two presumed psychogenic causes of chronic amnesia, namely, psychological illness and severe emotional trauma, the latter is the potentially relevant one in Neil's case. The many physical handicaps and neuroendocrine disorders from which Neil suffers could conceivably have triggered an emotional crisis, and this, in turn, might have led to a psychological breakdown resulting in some or all of the cognitive disturbances he displays. In fact, however, no psychological crisis was ever reported for Neil. Rather, the evidence points instead to a gradual alteration in his personality and emotional state accompanied by a similarly gradual deterioration in his perceptual and memory abilities. Such a progression seems more in keeping with progressive brain dysfunction induced by the tumour and its treatment than it does with an acute-onset, emotional trauma.

Despite the evidence against them, as well as the absence of any direct evidence for them, in the end neither a feigned nor a chronic functional amnesia can be decisively ruled out as an explanation for Neil's puzzling syndrome. However, an explanation in terms of a particular, though currently unknown, pattern of neuropathology also cannot be firmly ruled out. Additionally, in support of the latter possibility, there is the direct evidence that Neil did sustain diffuse neuropathology. Possible correlations between the brain-imaging findings and Neil's behavioural impairments are briefly reviewed next, following which his neuropsychological profile will be considered in greater detail.

Some neuropathological correlates

The findings in the CT and MRI scans taken of Neil before and after treatment (*see* Fig. 1) appear to explain his optic and motor disturbances, and also perhaps his amnesia. The scans performed at the time of his diagnosis revealed the

following: marked oedema of the midbrain and right cerebellum, which likely accounts for his ataxia and oculomotor difficulties; the cystic mass extending into the suprasella cistern, as well as the hydrocephalus, accounting presumably for the pituitary disturbances, the papilloedema, and the primary visual disabilities; and abnormal signal throughout the medial diencephalon and the white matter in the medial halves of both frontal lobes, consistent with his dense and global anterograde amnesia as well as with his personality change (Mair *et al.*, 1979; Victor *et al.*, 1989). None of the findings, however, appear to explain his cognitive as distinct from his primary visual difficulties.

By the time of the post-treatment scans, the tumour and accompanying oedema had largely resolved, leaving as residual abnormalities only some thinning or slight atrophy of many of the structures that had been involved initially. Yet none of the neurological or psychological difficulties listed above had resolved, except perhaps for the improvement in oculomotor control. Indeed, as already described, during the 18 months following treatment, his performance IQ (critically dependent on visual gnosis), memory and reading ability all deteriorated sharply in the absence of any evidence of new pathology. A detailed re-examination of the final, post-irradiation scans did reveal a region of possible atrophy involving the cortex and underlying white matter of the right inferior parietal lobule (*see* Fig. 2); however, given the location of the atrophy (as well as its ambiguity), it is unlikely to account for Neil's widespread cognitive deterioration (*see*, for example, Damasio and Damasio, 1983; Bauer and Rubens, 1985). The only explanation for this deterioration that can be proposed is delayed but undisclosed tissue damage resulting either from the initial oedema, the intensive radiation treatment, or both. Finally, nothing in the scans can begin to explain the unprecedented dissociation between Neil's amnesia and his ability to retrieve memories through writing.

Each of the foregoing issues will be considered again in connection with the analysis of his cognitive deficits.

Visual agnosia

Neil's severe impairment in identifying visual objects and photographs was confirmed repeatedly, both through informal observation of his behaviour and by formal testing (namely, with common objects, the Boston Naming Test, photographs of common household objects, facial expressions, face matching). This visual perceptual impairment must be largely responsible for his scoring in the mentally deficient range on the Performance Scale of the IQ test, a score that is out of keeping with his oral and written language and other intellectual skills (e.g. his successful performance on the Tower-of-London test) as well as with his verbal IQ, which falls within the average range. Neil's visual recognition failure extends even to his own creative drawings of objects and scenes (e.g. Fig. 3), unless he is asked to identify their contents by tracing them. Two recent adult case reports (Behrmann *et al.*, 1992; Jankowiak *et al.*, 1992) have documented that visual imagery may remain fully intact in the face of a severe visual agnosia. Rich imagery in the first of these agnosic patients enabled him to draw objects from memory in considerable detail, drawings that he later could not identify unless allowed to trace them. In these respects, Neil's case is identical and leads to the same conclusion, namely, that the central representations utilized for the recognition of familiar objects (an ability impaired in Neil) and the central representations utilized for the reproductive recall of those same objects (an ability spared in Neil) are normally either stored in separate locations or, more probably, retrieved from the same store through different pathways or different mechanisms.

Although Neil's visual sensory processes (visual fields, acuity, depth and colour vision, etc.) are seriously impaired, his perceptual failures are not due simply to these primary defects. This was also confirmed repeatedly in formal testing on certain of the perceptual tasks. His high scores on these tests (e.g. Tower of London, three-dimensional shapes, line orientation, Rey–Osterreith complex figure, Corsi Blocks), as well as his drawing skills, reinforce an earlier conclusion based on his performance on the Block Design Subtest of the WISC-R that his visuospatial perception is relatively intact. Preserved visuospatial functions possibly explain the two exceptions to his impairment in object perception that were noted earlier, namely, his fairly accurate identification of familiar objects in their familiar locations and of familiar photographs in a family album. Object familiarity, *per se*, seems insufficient as an explanation, since Neil failed to identify other highly familiar objects and pictured individuals outside their customary settings. Rather, in both successful instances, spatial cues may have substituted for object cues, a substitution he may well have been unaware of, just as he was unaware that tracing provided him with substitute kinaesthetic cues. (The use of substitute kinaesthetic cues in cases of visual agnosia was described as early as 1918 by Gelb and Goldstein; *see* Bauer and Rubens, 1985.)

If the foregoing interpretations are correct, then Neil's otherwise consistent failures in object and face perception reflect a severe but selective processing failure in object vision as distinct from spatial vision (for review, *see* Ungerleider and Mishkin, 1982), leading, in classical terminology, to a dense apperceptive visual object agnosia (Farah, 1990). According to current views, such an agnosia in the presence of preserved visuospatial perception implies selective dysfunction of an occipitotemporal as opposed to an occipitoparietal visual processing stream. As indicated above, the brain-imaging findings provide no support for this prediction in Neil's case, and so his visual agnosia, if such it is, remains unexplained other than by the possibility that the oedema associated with the malignancy led to undisclosed damage of the cortex or white matter in the occipitotemporal region.

Alexia

Neil's processing failure in object vision extends to words and letters, resulting in a dense alexia as well. His reading

disability was not so complete initially, however. At 4 months post-radiation treatment, his reading age was just 3 years below his chronological age; a year later it was about 9 years below; and not until 4 months after that did the alexia become complete. This is Neil's one cognitive skill for which there is quantitative evidence of a gradual decline across three different testing sessions. As indicated earlier, however, quantitative evidence was obtained across two of these sessions of a decline in both performance IQ and memory function.

It should be noted that cranial irradiation in the extensive form sustained by Neil often leads to delayed cognitive (and also endocrine) dysfunction, the effect not becoming manifest until a year or more following the treatment (Silverman *et al.*, 1984; Duffner *et al.*, 1985; Williams and Davis, 1986). The degree of loss is related to age at irradiation, the effect becoming milder the older the child (Danoff *et al.*, 1982; Chin and Maruyama, 1984; Siverman *et al.*, 1984). Also, when deterioration does take place, it ordinarily generalizes to all cognitive functions, including verbal IQ (Cousens *et al.*, 1988; Dennis *et al.*, 1992). Nevertheless, despite Neil's relatively late age at irradiation and the preservation of his verbal IQ, the progressive deterioration of his reading, as well of his other visual perceptual and memory abilities, are probably best explained as delayed effects of the radiation treatment superimposed upon the initial forebrain, midbrain and cerebellar damage.

Amnesia

Neil's intact remote memory has already been noted, this being a common feature, in fact a defining one, in cases of global anterograde amnesia. Whether he suffers from any retrograde memory loss, as amnesic patients commonly do for a period of up to a year or so preceding the onset of the causative lesions (Squire, 1987), cannot be known in his case because of the uncertain date of the responsible neuropathology.

Another sphere of memory that is commonly preserved in amnesic cases, namely, immediate memory, is not so highly preserved in Neil's case, at least not consistently. Thus, although he had an intact memory span on the Corsi blocks, he did not on digits. Similarly, whereas matching of successively presented wire shapes was intact with his right hand, it was not with his left. And again, while he could recall double consonants after unfilled delays, and showed a pronounced recency effect in auditory memory for word lists, he was unable to match successively presented melodies. Finally, although his copying of complex figures was highly proficient (e.g. Fig. 5B), his immediate reproduction of even simple geometric designs was equivalent to that of a 6-year-old (*see* Table 3). These mixed results on various measures of immediate memory do not allow a simple conclusion. His difficulties in this memory sphere appeared mainly with novel material such as the abstract wire shapes, unfamiliar melodies and visual designs, all stimuli for which he did not have pre-existing central representations, though this of course cannot account for his difficulties with lists of digits. With this single exception, however, it seems that Neil's immediate memory for familiar material is relatively preserved, a conclusion that is strongly supported by his facility with verbal exchanges and ability to follow verbal instructions.

In all other spheres of cognitive memory, Neil was profoundly impaired, consistent with the diagnosis of a dense and global anterograde amnesia. The impairment was evident in all aspects of his daily life as well as in all the formal testing we conducted, encompassing memory for biographical events from age 13 years onwards, orientation in time and retention beyond immediate memory of any verbal or non-verbal material presented through either the auditory, tactile or visual modality. This profound, global amnesia was also reflected in his severely deficient performance on the WMS, his memory quotient falling a full 50 points below his verbal IQ, which remained about average. Other evidence of his preserved intellectual status is provided by his skill in conversation, composition and drawing, as well as his satisfactory progress in school, as measured by his written work (*see below*). As proposed earlier, this profile of a dense and global anterograde amnesia in the presence of normal verbal intelligence may possibly be accounted for by the abnormalities observed on the brain scans in the area of the medial diencephalon and medial halves of both frontal lobes.

Memory retrieval through writing

The syndrome of anterograde amnesia, coupled as it usually is with preservation of both remote and immediate memory, is now commonly attributed to a defect in the mechanism responsible for placing newly acquired information into a long-term store. The same interpretation cannot be applied without modification to Neil's otherwise similar disorder in view of his remarkable ability to retrieve postmorbid memories through the act of writing, even while he is unable to give an oral account of the contents of his writing. It must be emphasized that these contents do not match those of his premorbid memory in either quantity or quality. His written answers to questions about his postmorbid experiences never contained more than a few items each or any richness of detail. Also, although his written digit span exceeded his oral digit span, his written recall of a word list did not surpass his oral recall. Neil's ability to recover postmorbid memories orthographically thus seems to be limited in generality as well as capacity, applying less to meaningless test stimuli than to meaningful experiences and events.

Unprecedented though it is, the dissociation in Neil's memory performance raises the possibility either that there are normally two different long-term stores for the same information or that the information in any given long-term store can be retrieved through two different response modalities. The results of two recent studies have provided strong evidence for the existence of two separate stores

and/or two separate retrieval modalities for verbal material. In one of these studies, an experiment by Tattersall and Broadbent (1991), normal subjects were required to recall lists of numbers by writing and lists of letters by speaking, or the reverse. Subjects who were told in advance which type of material was to be recalled orally and which type in writing performed significantly better than those who were not so instructed. Knowing the output modality in advance apparently permitted subjects to encode the material appropriately in preparation for recall, thereby implying the existence of two different lexical systems, one for oral and the other for written report. The second study, this one by Caramazza and Hillis (1991), contrasts the verbal impairments in two patients with left hemisphere strokes, one (H.W.) with damage to the parietal region and the other (S.J.D.) with damage to the fronto-temporal region. Both patients had greater difficulty in the production of verbs than of nouns, but case H.W. exhibited this difficulty only in speaking and not in writing, whereas the opposite was true for case S.J.D. This result likewise implies that there are two different lexical systems, one for articulatory and the other for orthographic output.

Although limited to the verbal domain, the findings in these two studies give added credence to the remarkable dissociation in memory ability exhibited by Neil. However, the problem for interpretation that his memory dissociation presents extends well beyond the verbal domain; i.e. he can retrieve through writing not only new information presented verbally, as in school work, but also day-to-day events that are ordinarily processed non-verbally. The evidence leads to the uncomfortable proposal that, in Neil's case, even non-verbal perceptions are nonetheless encoded and stored exclusively in an orthographic representational system or, alternatively, are stored in multiple representational systems serving memory for different types of sensory material yet are nonetheless retrievable exclusively through orthographic output. Neither alternative of course commends itself to common sense; yet, if the memory dissociation is genuine, no more acceptable alternative seems to be available. The selective anatomical disconnection implied by the above proposal is similar in principle to the one implied by Neil's short-term recognition of tactile shapes presented to the right hand but not the left, even though his identification of familiar objects with this hand is perfectly intact (*see* Tactile memory). Presumably, his left-hand tactual processing system, but not his right, is disconnected from even a short-term memory system; in this instance the disconection could have resulted in part from the damage to the rostral half of the corpus callosum that is evident on the MRI scans.

It is conceivable that Neil's ability to retrieve memories through writing is aided in some way by his intact kinaesthetic sense, but if so, the mechanism for it is unknown. He did not make use of kinaesthesis as a cognitive aid either before or during writing, but only afterward, when, in order to comprehend what he had written, he would slowly retrace his words, letter by letter. As indicated earlier, he could use kinaesthesis as a substitute for vision not only in reading, but also in object and pattern perception, yet he did not fully understand how this method worked, and he rarely employed it to decipher his own writing unless prompted to do so. As a result, he was most often completely unaware of what he had written just moments before.

The latter feature of Neil's syndrome is, of course, largely a product of his combined amnesia and alexia. The mystery is that in retrieving postmorbid memories orthographically, he is also totally unable to state orally what he is about to write as well as what he is currently writing. In this respect, Neil's orthographic performance resembles 'automatic writing', a dissociative phenomenon that was once the subject of intensive study in both experimental and clinical psychology (for an historical review, *see* Koutstaal, 1992). Perhaps the closest analogue to the observations reported here were instances of automatic writing by two female neuropsychiatric patients that were described recently by Joseph (1986). Both patients experienced repetitive bouts in which they produced large amounts of complex textual material of which they, too, remained unaware. These bouts, however, were sudden, non-volitional and seemingly paroxysmal, and were ascribed to the temporal-lobe epilepsy combined with the affective disorder from which each of them suffered.

As already indicated, there appears to have been no previous report of a case like Neil's in which an otherwise amnesic individual retrieved postmorbid memories by writing about them on request. It bears repeating that this ability was discovered accidentally by both examiner and patient. Our hope in presenting Neil's case is that other investigators of amnesia, being aware of the possibility of this phenomenon, might thereby be encouraged to search for it in their amnesic patients. Only if other instances of the phenomenon are encountered would it be profitable to speculate further about mechanisms that could give rise to so strange an entity as the dissociated form of episodic memory exhibited by Neil.

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Appendix

Neil's written answers to questions

8.3.90. The day it was discovered that Neil could retrieve memories through writing

1. Q: Do you remember anything about the book you are studying at school?
   A: Bloodshot Geranium windows Cider with Rosie Draniam smell of damp peppar and mushroomgrowth.
2. Q: Can you remember anything about what happened when you went to the hospital?
   A: A man had Gangreen.
3. Q: Who were some of your doctors?
   A: Dr. [name of endocrinologist] Dr. [name of neurologist] Dr. [name of oncologist] Dr. [First name of Registrar] Dr. [name of Senior Registrar].
4. Q: Who were some other people in hospital?
   A: [Name of tutor] [names of several other young patients on the ward] [name of play activity leader].

23.6.90. The day examiners took Neil on an afternoon outing, following a morning excursion with his father to an RAF display

5. Q: Who was at home when we left?
   A: My Mum, dad, Grandmother, and my sisters.
6. Q: What did we do this afternoon? Can you tell me?
   A: We went to [name of museum], and we had some pizza. Then we came back, we went onto the Beach and we looked at the sea. Then we came home.

(Questions Neil was asked to answer orally about the afternoon outing; answers in parentheses.)

(i) Did we go to a movie? (No).
(ii) Did we go to a restaurant? (No).
(iii) Did we walk on the board walk along the sea? (No).
(iv) Did we drive to [name of nearby town]? (No).
(v) Did we go to the . . . [local landmark]? (May be Yes . . . my . . . I just have a slight feeling inside me).
(vi) Did you have anything to eat? (No I don't think so).
(vii) Did we ask you any questions while we were driving? (I don't remember you doing so, but I would not be surprised if you did.)
(viii) Did we play games with you? (No).
(ix) Did we read you any lists of words? (No).
(x) Did you play a game in which you had to feel things with your hand? (I have a slight feeling that maybe).
(xi) Did we go to the beach behind your house? (No).

7. Q: How should my name be written?
   A: Dr. Kadem.
8. Q: What is my first name?
   A: Faraney.
9. Q: What did you have for lunch earlier today?
   A: I had a bacon sandwich.
10. Q: What did you do early this morning?
    A: I went to [name of town] to an RAF display in the [name of park]. I bougt one pound worth of posters, and I got two stickers for [names of sisters].

11. Q: What did you see while you were out at the RAF display?
    A: We sat in a raf tent and we looked at the life rafts and the new anti-aircraft guns.
12. Q: Did your mother play a game with you today?
    A: Yes.
13. Q: What was the game?
    A: She played a memory, remembering game. She was asking me about where I had been, who is in the house.
14. Q: Did your mother buy you something this morning?
    A: She went to buy (tight scribble) some (tight scribble) it was a (tight scribble).
15. Q: (Not recorded)
    A: I first discovered that I could use my hand at the Wolfson Center. With my my Mum and Dr. Cadem. I was sitting at the table copying out of a newspaper.
16. Q: How do you write Khadem?
    A: With a Kadem K.
17. Q: What did you do this morning?
    A: I got some sweets for [name of sister] this morning.
18. Q: (Not recorded)
    A: Dr. Elisabeth Issacs Dr. Cadem.
19. Q: Do you remember the first time you could read by copying?
    A: I was sitting at the table in front of a serving hatch in the wall. I copied large print from the newspaper. I was in the dining and the sitting room. We were looking onto an Astroturf football pitch.
20. Q: What did you have for lunch yesterday?
    A: spaghetti bolognaise.
21. Q: What was the first thing we did when we came into your house today?
    A: We went into my sisters bedroom.
22. Q: What did we do after that?
    A: We had some cake and the doctor was going to go. But he decided to come to see me.
23. Q: What will you do during your holidays?
    A: During the holidays Mr. [name of teacher] will come and teach me a new route.
24. Q: What did you do on your Sports Day?
    A: In my sports day I played bowles with sofballs.
25. Q: (Not recorded)
    A: During Art, wed am going bowling with the unit.
26. Q: (Not recorded)
    A: Mrs. [name of teacher] spoke to me about seeing the work experience officer.
27. Q: Who came with me to see you today?
    A: Dr. Michcin. he came from lake michegan in america.
28. Q: (Not recorded)
    A: I have entered for 100m walk. also games and rounders. It will be at a sports center in [name of village].

24.6.90. From Neil's mother's letter to Dr Khadem

I do hope you enjoyed our part of the world and had a safe journey home. When I got up this morning I questioned Neil on yesterday's events (asking for oral answers)—First I asked him what Day is it? Which he didn't know. Then I asked him what month? He said May. I corrected him and then asked him what month again. 5 mins later he said May. I also asked him what year is it he said 89. Then I asked him what did you do yesterday. First in the morning and then in the afternoon. He had a very *full memorable* day. He just sat and thought and couldn't answer me. In the end I frustrated him so much he burst into tears and walked out. He hasn't mentioned you what so ever! He has been to the shops this morning to buy 2 tomatoes, bananas and cream which they were out of. He went with [name of sister]. He also went to Swimming Pool place to by some chemical for swimming pool. Then we are going to have a barbecue for lunch—Chicken & Sausages, Garlic Bread and Salad.)

(The following questions were asked by Neil's mother.)

29. Q: Where did you go with Dad yesterday?
    A: I went to an R.A.F. display in [name of town]. I saw some liferafts which were inflatable, a plane, and other displays of ammunision.
30. Q: What month and year is it?
    A: The date is 28 [*sic*] June, 1990. The day is Sunday.
31. Q: Who came yesterday and where did you go?
    A: Dr. Kadem came yesterday with a doctor from America. We went to the [name of museum] and to have a snack. Then we walked in the lanes and when we got back we saw the sea.
32. Q: Where did you go with Dr. Khadem the day before (Friday)?
    A: On Friday Dr. Kadem and I went to the [name of hotel]. We played several games including 'The tower of Hanoi'.
33. Q: Yesterday where else did you go?
    A: After we went to the museum we had a snack in an italian restaurant.
34. Q: Did you eat and drink? What?
    A: I had a pizza and Dr. Kadem had a salad. I could not see the name. lemonade.
35. Q: What did Dr. Khadem buy me?
    A: She brought you a bunch of flowers.
36. Q: This morning what did you get from the shop? Who did you take?
    A: I went to the shops with [name of sister]. I bought 2 tomatoes and a lettece, and 1 packet of choclate, rasins for [name of other sister] and a chomp bar for [name of first sister]. You asked me to get some single cream.
37. Q: Where did you go with Dad? What did you get?
    A: I went to a garden center with dad, we got some swimming pool chemicals. They also had some rolls of selotape for sale.
38. Q: What tune did you like on the way home?
    A: On the way home I heard the Beatles song (Mother says this should have been The Nutcracker Suite).
39. Q: Did you walk in [name of town]? What else did you show her?
    A: She enjoyed the whole visit.
40. Q: Yesterday I was playing a game hiding something in my hand. What was it?
    A: Yesterday I was playing a game with mum. It was she was hiding a piece of string.
41. Q: What are we going to have for lunch today?
    A: A bar-b-que.
42. Q: What did you do in school today?
    A: Today at school I watched the film my left foot. I also had

geography. I saw Mr. (name of teacher) and he was very pleased. Miss (name of teacher) is going to give us a note about activities in the holidays.

13.7.90. Day of testing at the Wolfson Centre

43. Q: What did we just do?
    A: We just went upstairs and we went into a room with a blind. We did my visual fields and letter seeing. I afterward came down in the lift with Dr. Issacs.
44. Q: What do you have to do on this test you are doing now?
    A: I have to feel objects made of wire and say if they are the same.
45. Q: What books have you read (listened to on tape) recently?
    A: I have read Christie Brown which is another autobiography. I am now doing Julius Caesar which is Shakespear.
46. Q: What is the book you are currently studying at school?
    A: It is called My left foot.

2.8.90. Another day of testing at the Wolfson Centre

47. Q: Describe the test (tactile perception of wire shapes with right hand) you did before lunch.
    A: I had to wear a blindfold, and feel a twisted piece of coathanger. After I had finished I had to feel another piece and say whether they were the same, or different.
48. Q: Write with your left hand about the first test (memory of melodies) you did this morning.
    A: (Answered orally: Nothing is coming.)
49. Q: (Same as Question 48, but requested that he write answer with his right hand.)
    A: I had to listen to some music on a pair of headphones and say if I had heard it before.

3.8.90. A day of testing at Neil's home

50. Q: (Unrecorded)
    A: I copied a star onto a metal board, and I saw is reflection in a mirror. Copy a star in a mirror.
51. Q: (Unrecorded)
    A: I had to write what I had done previously.

4.8.90. Another day of testing, the last one, at Neil's home, following a visit on the previous evening by Dr Khadem

52. Q: What are these objects? (Six were shown in succession.)
    A: video casette
    [sea]shell
    green and transparent [pencil].

(Neil failed to identify a doll's comb, an ashtray, and a pencil case.)

53. Q: What were the objects I showed you earlier (asked after a 90-minute delay)?
    A: An ashtray A pencil with different colors The first thing that you showed me was a . . .
54. Q: (Unrecorded)
    A: I like Phil Collins. I don't know what instrument he plays.
55. Q: (Unrecorded)
    A: I wrote that I wanted a personal stereo.

(At examiner's request, Neil wrote answers to questions 56–83 with his left hand.)

56. Q: Did you go on an outing yesterday?
    A: I went to [name of the village].
57. Q: When did you get back?
    A: I got back about 7.00.
58. Q: Why were you delayed?
    A: Our coach broke down
59. Q: What did you have for dinner last night?
    A: Last night I had mince and potatoes
60. Q: How did your mother prepare the dinner?
    A: She barbecued.
61. Q: Who came to your home last night?
    A: You came last night.
62. Q: What time did I arrive?
    A: You came at approximately 8.30.
63. Q: Where were you sitting when I came?
    A: I was sitting at my desk.
64. Q: Where would that be?
    A: In the wooden chair.
65. Q: What did we do together?
    A: We used a new machine on which I drew the star.
66. Q: Had you seen it before?
    A: Not before Friday.
67. Q: Who was here besides us?
    A: Exept you there was my mother and father and my nan.
68. Q: Were they here when you and I were working?
    A: They went for a walk.
69. Q: So where did they go after?
    A: They went for a drink.
70. Q: Did I ask to use something in the house?
    A: You asked to borrow a pen.
71. Q: Where did I take it from?
    A: You took the pen from the holder on my desk.
72. Q: Did I ask to make a telephone call?
    A: You did but then you used your own phone.
73. Q: Where did I keep it?
    A: You kept your phone in your bag.
74. Q: What colour was it?
    A: I think it was white. (It was black.)
75. Q: Who saw me to the door before I left?
    A: I came to the door with you.

76–8. Q: Where did you go this morning? What for? Where did you go this afternoon?
    A: This morning I went [name of town] to get a personal stereo. I went to dions and Currys.

79–80. Q: What was it that I carried in my bag and did I leave behind anything?
    A: You left the star drawing machine behind. You carried the machine.

81–3. Q: On Friday in London, where did you have your lunch? What did you have for lunch? Did someone come to talk to you while you had your lunch?

A: I had my lunch in the park I had crisps sandwhiches and a drink. During my lunch you came to see me.

(At examiner's request, Neil wrote the remaining answers with his right hand.)

84–5. Q: Who sat next to you on the coach when you went on your outing, and what were the two things you brought with you to the Wolfson Centre on Friday?

A: On the coach to [Name of town] Stawart Mrs. [teacher's name] nephew sat next to me. On Friday I brought my stick with me. (Neil failed to write down the second item.)

86–7. Q: Where did you take Dr. Mishkin and me when we came to see you last, and what did you find in your car just as you were leaving the Wolfson Centre?

A: We went to [name of city] to the [name of museum] and afterwards we went to an Italian Restaurant. After we had been at the Wolfson Center I found a (tight scribble).

88–90. Q: What is the name of your doctor at the [name of hospital], the game that Dr. Isaacs and I bought you, and the name of the book that you recently read?

A: Dr. [name of endocrinologist] Othello my lef foot.

91–3. Q: What is your favourite rock song? Who is your best friend at school? Where do you go most often when you go for a walk?

A: I mainly listen to phil collins I mainly walk down the shops. (After a one-minute delay and a reminder that there was third question.) That was my best friend at school the person is [name of female friend], but she has left this term.

94. Q: (Unrecorded, following a session in which Neil had been taught to perform a series of hand movements.)

A: You showed me a sequence with your arms. left. on the settee. The movements are simular to semafore.