

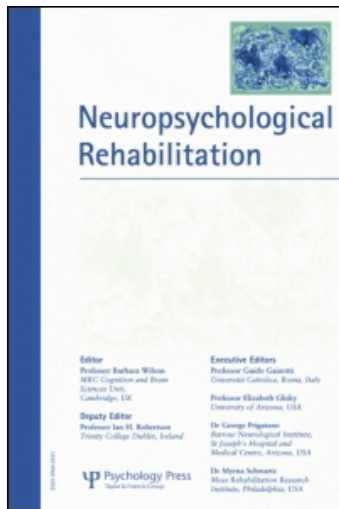
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Prominent and persistent loss of past awareness in amnesia: Delusion, impaired consciousness or coping strategy?

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Profound loss of awareness for the past in amnesia has implications for our understanding of memory and belief systems, and how they may become disrupted in neurological conditions. We report the case of CW, a professional musician who became severely amnesic in 1985 following herpes simplex viral encephalitis (HSVE) at the age of 46 years. For many years CW stated several times a day that he had just woken up. He frequently wrote this in his diary too. When shown examples of his diary entries or videos of himself playing or conducting music, he recognised both his handwriting and himself on the video screen but stated vehemently that he “was not conscious then”. In a previous paper (Wilson, Baddeley, & Kapur 1995), it was suggested that this lack of awareness for the past was a delusion, defined as a strongly held belief in the face of contradictory evidence (rather than implying any kind of psychiatric disorder per se). As a contribution to the academic debate regarding theories of “self”, in the present paper we will review this explanation of CW’s state as it had been in those early years, and we will also consider two other possibilities – namely, that CW had suffered from a loss of “autobiographical self” or “extended consciousness” (see Damasio, 2000, pp. 198–199), and that his verbal reports simply reflected a form of coping strategy to help him deal with the limited evidence he had available in “declarative” memory.

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INTRODUCTION

Severe memory loss, as in the amnesic syndrome, is often associated with differing degrees of lost orientation for person, age, time or place, and with varying degrees of memory distortion. Complete loss of self-knowledge, including loss of personal identity, is extremely rare in neurological disease, and is usually associated with psychogenic memory loss (Kopelman, 2002). Lack of awareness for one's age may occur in some neurological conditions, especially in the acute stages of a brain insult (Zangwill, 1953). Loss of awareness for time and place are usually associated with the acute phases of a brain injury (High, Levin, & Gary, 1990), with severe amnesia, or with the more advanced stages of a primary degenerative dementia. Distorted memories may take two broad forms – first, flawed “episodic memories”, in which there is either distortion of events that have occurred, such that events that never occurred may be introduced or mislocation of a true event to a different time and place; and, secondly, flawed “semantic memories”, where there are distortions in belief and knowledge structures. Delusions that accompany marked memory impairment could be seen to fall into the latter category (Schacter, 1995; Schacter & Scarry, 2000).

Delusions are notoriously hard to define. One definition is that a delusion is a belief that is clearly false and that indicates an abnormality in the affected person's content of thought (Hahn, 2003). Perhaps a tighter definition is that delusions are (1) false beliefs, held as (2) absolute convictions, (3) not amenable to argument, (4) not culturally explicable, and that (5) they are often bizarre, and (6) usually preoccupying (Clare, 1976; Mullen, 1979), but each component of this definition is debatable.

Delusions are not confined to psychiatric disorder but can be seen quite commonly in people with neurological conditions. The delusions most commonly reported in neuropsychological literature on people with brain damage are the (relatively rare) mono-thematic delusions, such as: (1) the Capgras syndrome, a belief that an acquaintance, usually a spouse or a close family member, has been replaced by an imposter (Ramachandran & Blakeslee, 1998); (2) the Fregoli delusion: the belief that one or several unfamiliar people are really someone very familiar (Ellis, Whitley, & Luaute, 1994; Markova & Berrios, 1994; Wright, Young, & Hellawell, 1993); (3) reduplicative paramnesia: a belief that a place or location has been duplicated (Sellal, Fontaine, van der Linden, Rainville, & Labrecque, 1996); and (4) Cotard's syndrome: a belief that one is dead or does not exist or is putrefying or has lost internal organs (Pearn & Gardner-Thorpe, 2002). The first three of these examples are

delusional misidentifications, and all four are relatively rare. Much more common in neurological and neuropsychiatric practice are delusions of persecution and reference, delusions related to mood state (e.g., derogatory, grandiose), delusions of being burgled (especially in dementia), somatic and religious delusions, or “first-rank” delusions of control and interference with thoughts (e.g., Cutting, 1997; Kopelman, in press; Lishman, 1998; Schneider, 1959).

In contrast to a delusion, a hallucination is a false perception occurring without any identifiable external stimulus (Hahn, 2003). More specifically, hallucinations involve: (1) false perceptions arising in the absence of an external stimulus, (2) occurring simultaneously with normal perceptions, and (3) having “substantiality” (i.e., they appear vivid, “real”). Furthermore, (4) they appear to be located in external space (“outside the head”), and (5) they arise independently (i.e., they cannot be conjured up or dismissed at will) (Kopelman, 1994; Mullen, 1979). One way of classifying hallucinations is according to sensory modality. Auditory hallucinations occur when people hear voices, sounds, noises or music in the absence of an external stimulus. The hearing of voices is the most common auditory hallucination, observed in many psychiatric patients but also in neurological and neuropsychiatric disorders.

Consciousness can be defined in several ways: Damasio (2000) offered a simple definition when he stated that it is “. . .an organism’s awareness of its own self and surroundings” (p. 4). Cartledge (2001) suggested that, in addition to awareness of self and one’s surroundings, there should be an ability to respond to environmental factors. Gray (2004) suggested that, rather than trying to define consciousness, it may be more fruitful to consider the various distinctions that occur in discussions of conscious experience – he distinguished between conscious awareness of the outside world (“public cognitive space”), conscious awareness of one’s thoughts and images (“private cognitive space”), and awareness of inner bodily sensations, such as those of hunger or tiredness (“private bodily space”). In common with other authors (e.g., Zeman, 2002), he also argued that it is important to distinguish between the contents of conscious experience and states of consciousness – the latter usually referring to degrees of wakefulness.

We report on a very amnesic patient who, despite playing and conducting music and writing diary entries several times a day until approximately a year ago, consistently denied he was conscious at the time. Although this state persisted for many years, we have been unable to track down any reports of patients who have denied that they had ever been conscious since the onset of their illness, despite evidence to the contrary, for such long periods of time.

CASE REPORT

CW was born in 1938. He was an outstanding musician and gifted musical scholar, being one of the world’s experts on Orlando Lassus, the Renaissance

composer. He produced BBC Radio 3 programmes of Renaissance Royal wedding music to celebrate the wedding of Prince Charles and Lady Diana Spencer and was leader of the London Sinfonietta. In March 1985 at the age of 46 years, he developed an influenza type illness with headache and fever. He was admitted to hospital days later and the diagnosis of herpes simplex viral encephalitis (HSVE) was made. His level of consciousness fluctuated, and he was treated with acyclovir. This probably saved his life, but the virus destroyed large sections of his brain. (For a detailed account of the onset of his disorder, see Wearing, 2005, also Wilson & Wearing, 1995.)

An MRI scan was carried out in July 1991 (see Figure 1). Three independent raters agreed there were marked abnormalities bilaterally in the following areas: hippocampal formations, amygdalas, mamillary bodies, temporal poles, and substantia innominata. In addition there were marked abnormalities unilaterally (left) in the fornix, inferior temporal gyrus, anterior portion of middle temporal gyrus, anterior portion of superior temporal gyrus, and the insula. There were mild abnormalities in the posterior portion of left middle temporal gyrus, the left medial frontal cortex, the left striatum, the right insula, the right fornix and the anterior portion of right inferior temporal gyrus. In addition, the third ventricle and both lateral ventricles were significantly dilated. Both the left and the right thalami were considered to be intact and no other frontal abnormalities were seen other than the left medial frontal area mentioned above.

A second MRI (3-Tesla) was carried out in January 2006. As shown in Figure 2, there was very severe and extensive loss of left temporal lobe

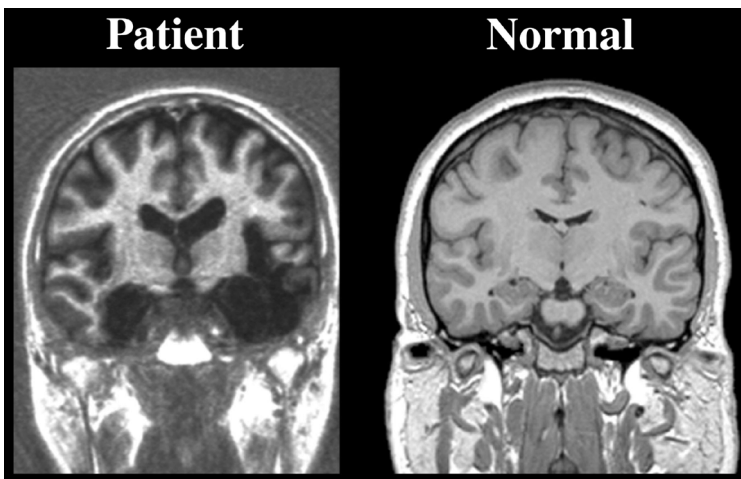


Figure 1. CW's MRI scan from 1991 compared with a non-brain damaged age-matched control.

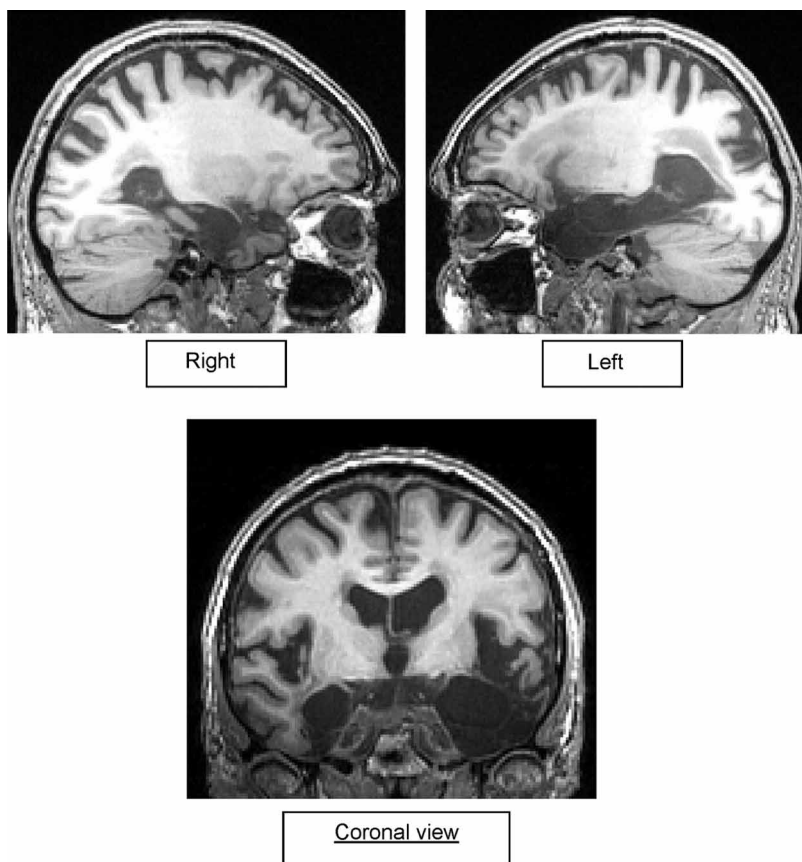


Figure 2. MRI 2006 showing that CW has extensive damage to the temporal cortices, greater on the left than on the right.

substance, and similarly severe loss of right medial temporal lobe tissue with some degree of generalised cortical atrophy throughout the brain. Comparing the 1991 and 2006 MRI scans, and allowing for improved imaging technology, there was very little change.

Neuropsychological assessment

CW was referred to one of us (BAW) in October 1985. A previous psychological report said there were extremely severe episodic memory deficits, some semantic memory impairments and CW's immediate memory span was normal.

A summary of the neuropsychological assessment of November 1985 can be seen in Table 1. In essence, CW's Verbal and Performance IQ scores were in the average range. This was, undoubtedly, a decline from his premorbid ability, which must have been in the superior or very superior range given his achievements. He showed evidence of executive dysfunction, particularly on a test of verbal fluency. As was the case in the earlier report (Wilson et al., 1995), his immediate memory, as measured by forward digit span (span of 7) and tapping a sequence of Corsi blocks (span of 5), was normal with delayed recall in the severely impaired range. Furthermore, CW, who had always been a city dweller, described a picture of a scarecrow as "a worshipping point for certain cultures" and, when asked, "What is a scarecrow?" he said, "A bird that flies and makes funny noises", which was interpreted as evidence of semantic memory impairment.

CW has been assessed on many occasions over the past 21 years. He has always scored zero on tests of delayed recall, and he has been unable to lay down new semantic information. In January 2006, on a test of new semantic knowledge, CW was given 50 words to define, which had come into the language over the past four decades (for example, e-mail, mad cow disease, and London Eye). The only word that CW defined accurately was "Eurotunnel", which could have been from guesswork. However, he has recently begun to show some evidence of new semantic learning, for

TABLE 1
Summary of CW's neuropsychological test scores

	1985	1992
•NART	122	111
•WAIS-R IQ	106	97
•WMS-R General index		<50
•Delayed index		<50
•RBMT	0/12	0/12
•Autobiog. Memory Interview		Severe impairment exc. childhood facts.
•Graded Naming		2/30
•Semantic Battery Naming		
–Animate		30/60
–Inanimate		52/60
•Frontal/Executive:		
–Card sorting categories	4/6	6/6
–perseverations	8	0

Wilson et al., 1985, 1992.

example, the name and use of a mobile phone (for other examples, see Wearing, 2005).

CW has shown some evidence of residual learning capability, e.g., he could readily navigate to parts of the residential home in which he was living, such as the kitchen, even though it was difficult for him to give an explicit description of the route from memory if asked to do so. Furthermore, when CW took part in an errorless learning study (Baddeley & Wilson, 1994), he showed a major benefit for error-free over errorful learning, with a total of 12/36 correct in the errorful condition and 22/36 correct in the errorless condition. We argued that the superiority of errorless learning was due to implicit memory mechanisms, and Page, Wilson, Shiel, Carter, and Norris (2006) gave further support for this view. On a more formal test of implicit memory (Nannery, Sopena, Greenfield, & Wilson, 2007), CW showed some savings on a test of stem completion and on a perceptual priming (fragmented pictures) test. We have been unable to establish whether CW can learn new musical pieces, but he never learned pieces before he became ill as he always sight-read and he can still do this.

CW suffered an extensive retrograde amnesia, with markedly impaired recall on the Autobiographical Memory Interview (Kopelman, Wilson, & Baddeley, 1989) for all time-periods. He could recall very few facts from before the illness. Although he knew who he was and that he was a well-known musician, where he went to school, where he studied music, and that he married his wife, details were very patchy and he had lost most of his episodic memories for the 45 years prior to his illness. On a test of famous faces and names, CW was shown 18 well-known people. He recognised none of them and said only four were vaguely familiar (for example, he thought Margaret Thatcher was vaguely familiar and that she was a member of the Royal family and that Prince Philip was a member of CW's choir). When given their names, he gave identifying information for two, and four others were described as "vaguely familiar".

In summary, CW's memory functioning, as assessed by conventional neuropsychological tests, remained largely unchanged over 21 years, although Wearing (2005) noted some changes in everyday memory and conversation consistent with his being able to hold information in his memory longer than he used to (for example, he can now follow the plots of films more easily).

CW's auditory hallucination

In 1990 CW developed an auditory hallucination. He thought he could hear his music being played and said to his wife, "Listen, they're playing my music again." His wife believed he could genuinely hear it as he would start to hum the tune and pick it up mid-phrase. She indicated that if he

were pretending to hear it then she thought he would start at the beginning of a phrase. This musical hallucination has remained and CW still “hears” his own music several times a week (described as “they’re playing one of my tapes”), although he does not experience any other auditory hallucinations. Evers and Elger (2004) reviewed published cases of musical hallucinations associated with psychiatric and neurological aetiologies. They pointed out that, as in the case of CW, most patients with musical hallucinations perceive familiar songs, often the same one played repetitively (as in CW’s case), implicating residual memory traces in the generation of the hallucinations. They also considered whether laterality of lesion was critical in generating musical hallucinations in neurological patients, and concluded that the small advantage for right-sided lesions “was not significant and supports the hypothesis that right-sided lesions play a role only marginally more important in the aetiology of musical hallucinations than left-sided lesions” (2004, p. 60). It thus remains possible that the origin of CW’s musical hallucinations were from the more extensive left temporal lobe lesion that was characteristic of his lesion profile. It should also be noted that musical hallucinations are often associated with temporal lobe epilepsy (Lishman, 1998), and CW has indeed experienced complex partial seizures secondary to his extensive temporal lobe pathology, and he remains on anti-convulsant medication.

CW’s belief that he was not previously conscious

For many years, CW did not appear to accept that he has a memory disorder, attributing his problems to the fact that he has not been conscious since he became ill. Indeed, one of CW’s most often repeated phrases was that he had become conscious for the first time. In the early years, he reported this many times a day and added, “it is like being dead”. A typical statement was: “I have just become conscious for the first time, this is the first sight I’ve had, the first taste I’ve had (sipping his coffee) it’s like being dead. Does anyone know what it’s like being dead? Answer, no”. Sometimes he spoke as if he *were* dead. For example, on one occasion he was talking about millionaires and said, “When I was young there was only one millionaire, *but when I died* there were millions.” If shown his diaries in which he had recorded his moments of just awakening, he accepted that the entries were in his handwriting but said, “But I wasn’t conscious when I wrote that, I’m now conscious for the first time.” If shown videotapes of himself playing the piano or conducting his choir (which he has done on occasion for television documentary programmes), he recognised himself in the video clips but, once again, said, “I wasn’t conscious then”. He did not accept that his problem was due to a memory impairment, but stated that it was due to a failure of consciousness since he became ill. Furthermore, if challenged about this belief he would become angry, because his subjective

experience of amnesia was as awakening from total void – it could not be explained, because there was no vocabulary for CW's amnesic window on the world. Although it would have been interesting to discuss with CW what he actually meant when he said that he was now conscious for the first time, this was impossible because, as soon as one tried to engage in such a discussion, CW would interrupt to say that he had just woken up. In-depth conversations were impossible because of (1) his dense amnesia and (2) his preoccupation of having just regained consciousness.

In 1995 Wilson et al. suggested that this could perhaps be interpreted as a delusion, although without any psychiatric connotations. We reported that he must have been conscious when he wrote in his diary, played the piano and conducted his choir. We thought then that there were two aspects to consciousness – *being awake* and *being aware* and, as CW was both awake and aware, he must have had conscious experience.

Was CW suffering from a delusion?

At one level, it is obvious that CW was conscious in order to make the statement that he had not been conscious before. One cannot make such a statement if one is unconscious, asleep or in a coma. He also had experienced a recurrent auditory hallucination so he displayed two forms of misinterpretation about himself and the world about him. Was his belief about lack of past consciousness (i.e., loss of consciousness since his illness for he did not deny that he was *ever* conscious) due to the location of his brain damage or to the severity of his brain damage? Following Jaspers' (1913) distinction between the "form" of mental phenomena (or psychopathology) and their (specific) "content", Feinberg, Eaton, Roane, and Giacino (1999) have argued that memory and executive deficits are a necessary (although insufficient) condition for the occurrence of delusions in neurological disorders (and in psychiatric disorders), but that their content can be influenced by motivational and other factors. CW certainly had severe memory deficits and some executive deficits, although, of course, not all patients with combined executive and memory disorders experience delusions.

What about the specific content of CW's delusion? It may simply be an attempt to explain to himself *why* he cannot remember, and the content of the auditory hallucination may also have been influenced by fact that he is a musician. However, we know of no other reports in the literature where people have so persistently and predominantly denied past consciousness, although similar much more transient complaints do occasionally occur in amnesic patients. Ramachandran and Blakeslee (1998) suggest that Capgras syndrome is a result of a disconnection between temporal lobe and limbic system. There is no evidence that CW currently has any other delusions,

either common ones, such as ideas of persecution or reference, or rare ones, such as the Capgras syndrome or Fregoli's syndrome.

Given the superficial resemblance between saying that one had not previously been conscious and saying that a body part or oneself is dead, it might appear pertinent to ask if CW had a variant of Cotard's syndrome. This does not seem to be the case, although he has occasionally spoken as if he had been dead (saying "after I died" instead of "after I became ill"), he was far more likely to say, "It is like being dead". In addition, he did not show other features of Cotard's syndrome such as suggesting that he did not exist or that he was putrefying or had lost his internal organs – moreover, unlike many Cotard patients, CW did not show any evidence of depression.

In brief, while the fixity of CW's belief is indeed consistent with its being *described* as "delusional", CW does not report other psychiatric features, which would suggest that an underlying "organic" psychosis is an *explanation* of his complaint of absent past consciousness. Thus, we have modified our original view and now no longer believe "a delusion" is a sufficient account of this phenomenon.

CW's belief as a form of impaired consciousness

Since our paper in 1995, much more has been written about consciousness. Zeman (2002), for example, in his excellent book reminded us of the complexity of consciousness and referred to scientific and philosophical theories of the phenomenon. In 2006, Zeman also wrote about self-consciousness in which he included self-perception, self-monitoring (metacognition), self-recognition (e.g., of body in mirror), awareness of awareness (possession of theory of mind) and self-knowledge (autonoetic awareness). Interesting though this topic is, it requires another paper to deal with it as it lies beyond the scope of this paper.

Damasio (2000) has argued that there are three types of consciousness or self. The most basic is proto self: "a coherent collection of neural patterns which map, moment by moment, the state of the physical structure of the organism in its many dimensions" (p. 154). Damasio (2000) went on to say that "we are not conscious of the proto self . . . [it] has no powers of perception and holds no knowledge" (p. 154). The next level is core self or core consciousness. This kind of self is condemned to endless and fruitless transiency. According to Damasio, all that core consciousness requires is a brief short-term memory, and we know that CW certainly had this. Then there is a higher form of consciousness that Damasio called autobiographical self, which is required for extended consciousness. Damasio wrote: "I use the term autobiographical memory to denote the organised record of the main aspects of an organism's biography." CW certainly had some knowledge of

his autobiographical memory but this was impoverished for the period before he had become ill and, since his illness, he had laid down virtually no new autobiographical facts. According to Damasio, autobiographical self is constantly being refashioned as a result of experience. He also wrote that, "Extended consciousness depends on holding in mind, over substantial periods of time, the multiple neural patterns which describe the autobiographical self" (p. 200). We suggest that CW could not hold in mind these patterns over extended periods of time.

Autobiographical self then is the state when certain records of one's personal past are made explicit in reconstructed images as needed. This would appear to be the kind of deficit suffered by CW. He could not reconstruct images from his autobiographical memory. He had a very severe autobiographical memory impairment both for episodes prior to his illness and for the years since his illness. He could not lay down any new memories. Damasio argued that being conscious goes beyond being awake and attentive, it requires an inner sense of self in the act of knowing. The primary damage to CW's brain lies in the limbic areas which, Damasio argues, allow him to have core consciousness but not autobiographical consciousness. This leads to a life "being sensed but not really examined" (p. 217) and this, in turn, could explain why it was so difficult to have an in-depth, meaningful conversation with CW.

In support of this view, Damasio described a patient of his, David, who was similar to CW in the extent, severity and location of his brain lesions. Although David did not say he had just woken up or that he had not been conscious before, he did say that Damasio was his cousin, suggesting the presence of a form of Fregoli illusion. The amount of hippocampal damage seen in David was similar to that seen in HM (Scoville & Milner, 1957) but, unlike HM, David had damage to the cortices of the temporal lobes especially in the inferotemporal and polar regions. CW has similar damage to David, i.e., both in the hippocampal areas and in the temporal cortices (see CW's scan in Figure 2). Damage to the temporal cortices, then, should not impair core consciousness, since the structures required for this are intact, but it will impair the activation of autobiographical memory records and thus reduce the scope of extended consciousness. Presumably, the severity and extent of CW's temporal lobe damage relates to his failure to recognise the existence of his past consciousness.

Was CW's belief simply a form of coping strategy?

In her description of CW's "moments of awakening" in the early stages of his illness, Wearing (2005) noted, "As it seemed impossible to fix anything in his mind, it was as if every waking moment was the first waking moment. Clive

was under the constant impression that he had just emerged from unconsciousness because he had no evidence in his own mind of ever being awake before.” (p. 127). If this belief were challenged, he would become irritable; and even if he were to be talked through the logic of what he was saying, he would rapidly return to this belief. Indeed, this irritability, in the absence of any overt depression or anxiety, was perhaps the most salient aspect of CW’s mood state.

It would appear that CW had little or no declarative memory support when faced with situations which he had to explain, whether it was entries in his diary, meeting someone again a few minutes later, etc. His coping strategy for the absence of such declarative memory appeared to be to generate false memories, or to deny any previous experience with the items in question. It would appear that for CW, “consciousness” and “awareness” were interchangeable, and that if he had no “memory”, and thus awareness, for immediately preceding events, he therefore believed that he was “aware” for the first time. He thus did not have the continual “stream of consciousness” or “stream of awareness” that Penfield eloquently referred to in his studies of patients with temporal lobe epilepsy (Penfield, 1959). CW’s repeated denials could be seen to be a form of coping strategy to deal with this complete discontinuity in his stream of consciousness. As Bogousslavsky and Inglin (2007) have pointed out, beliefs can be seen to serve two functions – to represent a concept, and to form a justification for a viewpoint or for a course of action. In CW’s case, his belief that he now had the impression of being conscious for the first time helped him to cope with his absent memory and also provided a justification for his subsequent statements and behaviours.

CONCLUSIONS

CW knew who was he but had limited knowledge about many aspects of his life. Klein, Rozendal, and Cosmides (2002) have argued that there are multiple contributions to the concept of self, including episodic memories, representations of one’s own personality traits, knowledge of the facts of one’s own life, the sense of continuity of experience through time, a sense of personal agency, and the ability to self-reflect on one’s own mental states. Seen in this context, CW’s sense of self had almost certainly been disrupted by his memory disorder. His persistent belief that he was “conscious for the first time” could be seen as a form of (isolated) delusion (without any psychiatric implication), or an impairment of consciousness, or a coping strategy to deal with the limited evidence that his cognitive deficits allowed him to have at his disposal. This belief no doubt fed into his disrupted sense of self, and combined to contribute to the personality and behaviour that he displayed in his

interactions with others. We should perhaps add that, in recent years, there have been some changes in CW's semantic learning, his emotional disposition, and his insight (see Wearing, 2005), which we plan to investigate further in future publications.

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