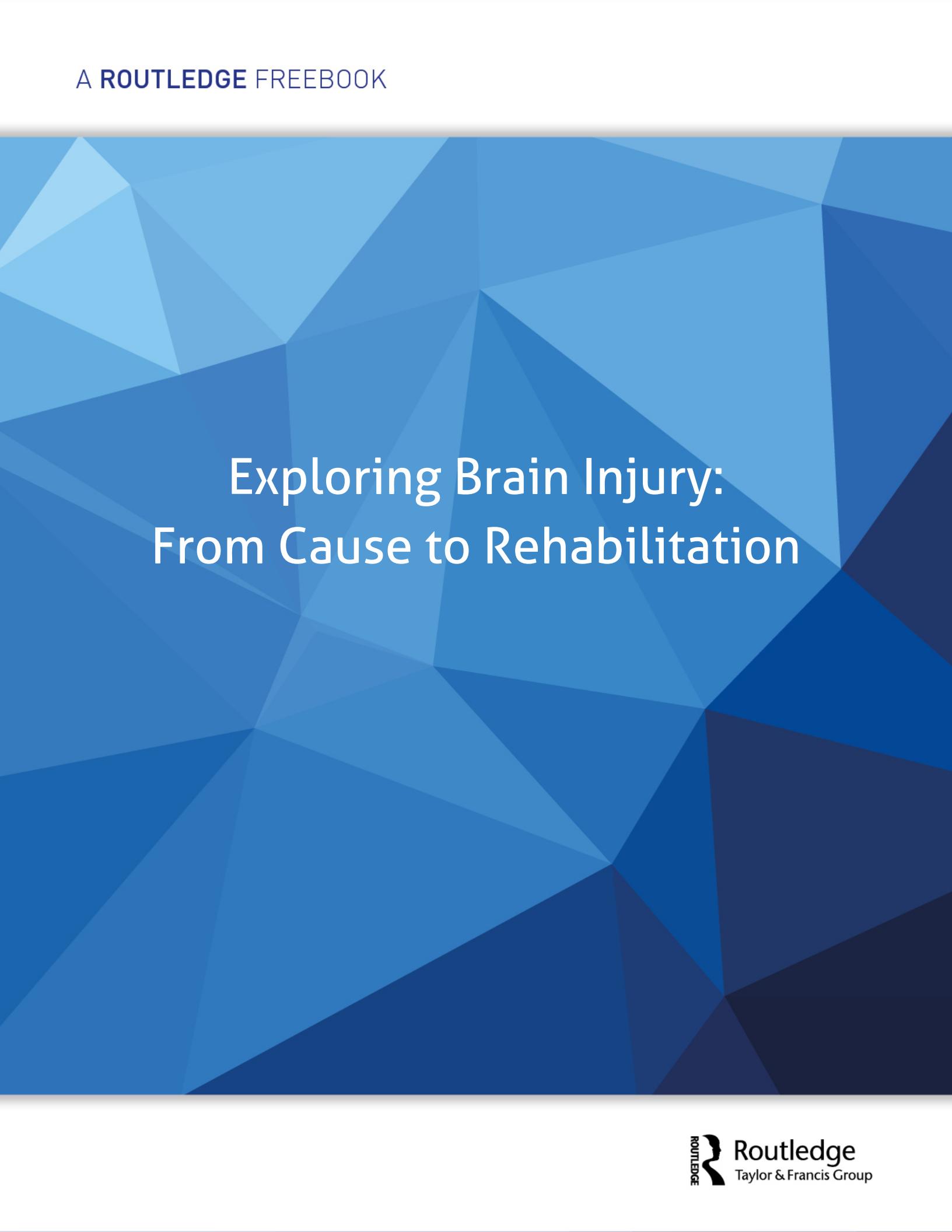


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The background of the cover features a complex, abstract geometric pattern composed of numerous triangles in various shades of blue, from light cyan to dark navy, creating a sense of depth and movement.

Exploring Brain Injury: From Cause to Rehabilitation



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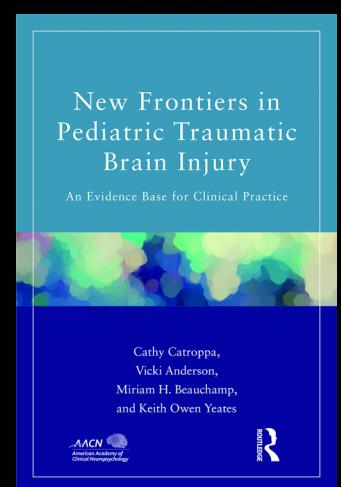
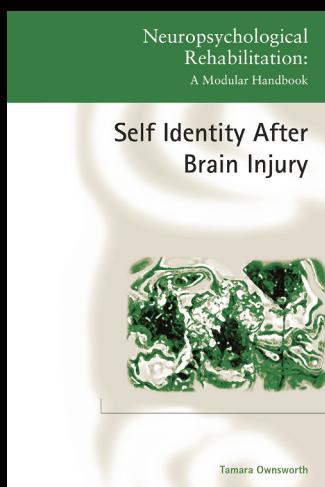
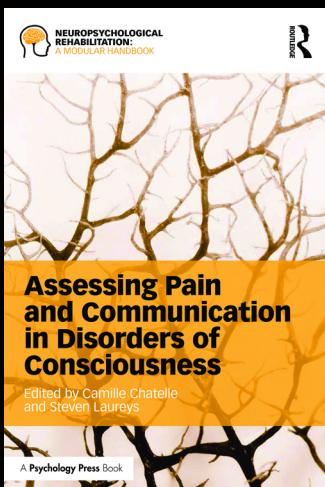
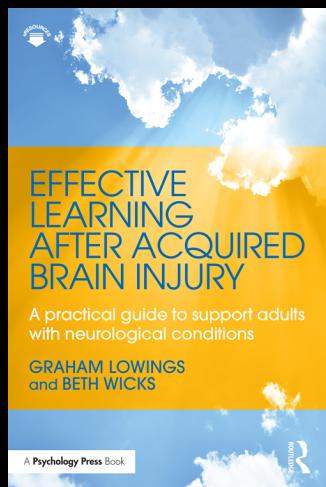
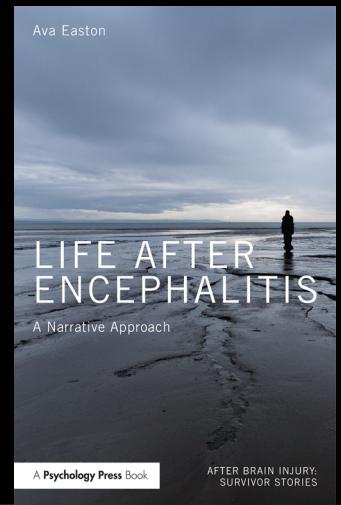
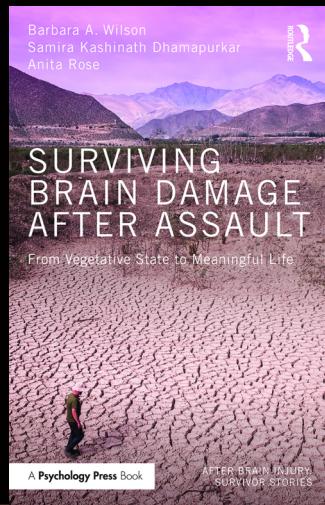
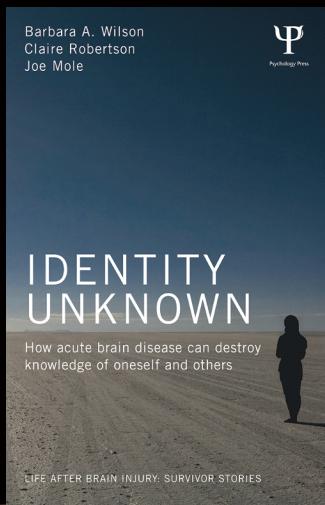
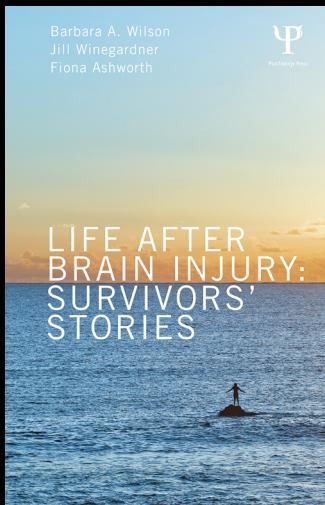
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Introduction

The World Health Organisation has recently acknowledged the need to raise the profile of mental health issues (with the WHO Mental Health Action Plan 2013-20) and we believe there needs to be a similar focus on psychological, neurological and behavioural consequences of brain disorder, and a deeper understanding of the role of rehabilitation in making the lives of brain injured people more tolerable. Giving a voice to survivors of brain injury is a step in the right direction.

This FreeBook is aimed at those who have suffered a brain injury, their families and carers, and professionals who are involved in neuropsychological rehabilitation. Brain disorders can be life-changing events with far-reaching consequences. However, in the current climate of cuts in funding and service provision for neuropsychological rehabilitation, there is a risk that people whose lives have been transformed by brain injury are left feeling isolated with little support. We hope to highlight the importance of raising awareness of those that have suffered a brain injury and the importance of rehabilitation in the recovery of sufferers.

Chapter 1

In this chapter from *Life After Brain Injury*, key members in the field, Barbara A. Wilson, Jill Winegardner and Fiona Ashworth, from the Oliver Zangwill Centre for Neuropsychological Rehabilitation, provide an overview of brain injury and the importance of rehabilitation in a sufferer's recovery.

Chapter 2

This introductory chapter highlights the importance of addressing self-identity issues in brain injury rehabilitation. Written by Tamara Ownsworth, a teaching and research academic in the School of Applied Psychology at Griffith University in Brisbane, Australia it also presents an overview of her book, *Self-Identity after Brain Injury*.

Chapter 3

Written by Camille Chatelle and Steven Laureys, this chapter examines the difference disorders of consciousness (DOC) associated with brain injury and the clinical challenges associated with these patients in terms of pain management, detection of consciousness and communication.

Chapter 4

Encephalitis, which causes inflammation of the brain, is a devastating condition whose impact upon people should not be underestimated. *Life After Encephalitis: A Narrative Approach* by Ava Easton, the CEO of The Encephalitis Society, provides a unique insight into an often misunderstood condition which robs people of abilities most of us take for granted.

Chapter 5

In this chapter from *Effective Learning after Acquired Brain Injury*, Graham Lowings and Beth Wicks, specialists in acquired brain injury, explain how to take the results of a neuropsychological assessment as a guide in order to construct a cognitive profile and to create individually tailored educational plans and rehabilitation programmes.

Chapter 6

Barbara A. Wilson, founder of the Oliver Zangwill Centre for Neuropsychological Rehabilitation, Claire Robertson, a sufferer of brain injury and Joe Mole, a psychology assistant at the Oliver Zangwill Centre for Neuropsychological Rehabilitation highlight strategies that helped the rehabilitation of a sufferer of a viral infection of the brain, and how they coped with the severe after-effects of brain damaged caused by it.

Chapter 7

Rehabilitation is extremely important and this chapter, from *Surviving Brain Damage After Assault*, by Barbara A. Wilson Samira Kashinath Dhamapurkar and Anita Rose portrays the remarkable recovery of Gary, who was assaulted by a gang with baseball bats and a hammer, resulting in several skull fractures and severe brain damage.

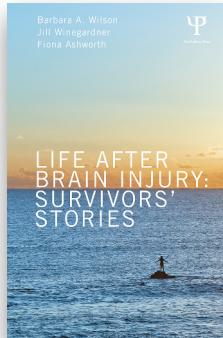
* As you read through this FreeBook, you will notice that some excerpts reference previous chapters—please note that these are references to the original text and not the FreeBook.



An Introduction

by Barbara Wilson

Chapter 1: An Introduction



The following is excerpted from *Life After Brain Injury* by Barbara A. Wilson and Jill Winegardner, Fiona Ashworth. © 2014 Taylor & Francis Group. All rights reserved.

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Unlike damage to a limb or indeed even another organ, damage to the brain can be debilitating to an extent that those injured remain dependent on others for the rest of their lives. Except in rare circumstances, a brain cannot be substituted by an artificial device or treated back to its original form or capacity, so brain injured people have to learn how to live their lives with handicaps that are not going to disappear over time.

In recent times workers in the field of neuro-rehabilitation have built up an extensive knowledge of the consequences of injury to the brain, they have developed sophisticated ways of analysing and diagnosing problems faced by brain injured people, and they have developed scientifically supported methods of treatment. The neuropsychologists who have contributed to this book have been able to draw upon an extensive literature of research and treatment as well as their own experiences in the field of brain injury rehabilitation. We stress again, however, that what makes this book unique is that the team of authors has been extended to include brain injured survivors themselves, thereby providing as full an account of brain injury and its consequences as possible. We are fairly confident that this book goes deeper and further than most if not all other books describing case studies dealing with the effects of brain injury, which rarely if ever include the story from the patient's point of view, and that barely touch upon the intricacies and efforts involved in brain injury rehabilitation.

There is much misunderstanding about the nature and consequences of brain injury and unfortunately this sometimes extends into the professional community and even exists among medical and political authorities who ought to know better! It is far too easy for some in authority, who do not wish to use some of the funds for which they are responsible, to accept that once a person can walk out of a hospital nothing else needs to be done. This is rarely the case for someone who has received an insult to the brain. Holders of the country's purse strings can easily be seduced by modern technology, preferring to spend money on machines that can light up colours in the brain but cannot as yet indicate treatment for the individuals whose brains light up! Meanwhile professionals working in neuro-rehabilitation are starved of funding despite the fact that brain injured people are convincingly helped to lead better daily lives after receiving treatment at the hands of clinical and neuropsychologists, occupational therapists and speech and language therapists. The irony is that brain injured people who do not receive the sort of rehabilitation described in this book can ultimately become a much larger financial burden upon the state. Rehabilitation not only makes life better for brain injured patients and their families, it also makes economic sense.

There is evidence to show that rehabilitation may be expensive in the short term but it is cost effective in the long term (Prigatano & Pliskin, 2002; Winegardner & Ashworth,

2012). There are several studies suggesting that rehabilitation for survivors of brain injury is economically effective. For example, Cope *et al.* (1991) surveyed 145 American patients and found that the estimated saving in care costs following rehabilitation for a person with severe brain injury was over £27,000 (\$40,500) per year. The number of people requiring 24 hours per day care dropped from 23% to 4% after rehabilitation. A Danish study (Mehlbye & Larsen, 1994) reported that expenditure in health and social care for patients attending a non-residential programme were recouped in five years. The costs of *not* rehabilitating people with brain injury are considerable given the fact that many are young with a relatively normal life expectancy (Greenwood & McMillan, 1993). Cope (1994) suggests that post-acute rehabilitation programmes can produce sufficient savings to justify their support on a cost–benefit basis.

On a slightly different theme, a study by West *et al.* (1991) claimed that people with traumatic brain injury (TBI) who had attended a supported work programme earned more than the programme costs after 58 weeks of supported employment. Furthermore, after two and a half years there was a net gain to the taxpayers who had ultimately funded the service. This did not include the indirect costs such as savings from family members who were able to return to work. Indirect costs have also been reported by Teasdale *et al.* (2009), who found that the strain on caregivers was reduced following use of a pager to remind patients what to do. Wood *et al.* (1999) wanted to establish the clinical and cost-effectiveness of a post-acute neurobehavioural community rehabilitation programme provided for 76 people surviving severe brain injury. The majority had sustained their injuries more than two years prior to admission and all had spent at least six months in rehabilitation. In terms of improved social outcomes and savings in care hours, it was found that the most cost-effective benefit was the provision of rehabilitation within two years of head injury; and it was still worthwhile, in terms of clinical and cost-effectiveness, to offer rehabilitation to those who were more than two years post-insult. Not only can rehabilitation save lives and improve quality of life for people with brain injury, it can also lead to major savings for government systems. It is estimated that people with moderate to severe brain injuries will have health and social care service costs of between £20,000 and £40,000 per year. A 25-year-old denied rehabilitation of the kind described in this book, and who lives to 75 years, may accrue costs of between £1,000,000 and £2,000,000.

There is plenty of evidence to show that comprehensive neuropsychological rehabilitation is clinically effective. Cicerone and his colleagues, for example, in a meta-analysis, found that such programmes can improve community integration, functional independence and productivity, even for patients who are many years post injury (Cicerone *et al.*, 2011). Van Heugten, Gregório and Wade (2012) looked at 95 randomised control trials carried out between 1980 and 2010 and concluded that there is a large body of evidence to support the efficacy of cognitive rehabilitation.

Rehabilitation for people with brain injury, conducted by qualified experts, is complex and demanding of expertise. It is not simply a tag on to medical treatment; neither is it in any way less complicated than analysing the results of MRI scans. We hope to show this in the following pages, and initially I would like to present a framework for rehabilitation that I developed a few years ago (Wilson, 2002) showing the areas that need to be considered and negotiated in order to provide effective treatment to brain injured individuals (see Figure 1.1). Obviously, not all areas will apply to each of the clients in this book. However, when contemplating each treatment described, the reader should be able to recognise particular pathways through the framework. At the end of this book we return to the framework to ask the reader whether we have kept to these pathways and been able to reach any kind of successful destinations as far as the clients are concerned.

The starting point for any rehabilitation programme is the patient or client and his or her family. In addition to background and ethnic and social issues, the nature, extent and severity of brain damage should be determined. Current problems, including cognitive, emotional, psycho-social and behavioural, need to be assessed. Models of language, reading, memory, executive functioning, attention and perception are available to provide detail about cognitive strengths and deficits. We can use assessment tools to determine emotional, behavioural, and social difficulties. Behavioural or functional assessments can be used to complement standardised assessment procedures.

Having identified problems, the rehabilitation programme can be planned. Patients, families, and staff need to negotiate meaningful, functionally relevant, and attainable goals. Ways to achieve these goals need to be formulated (for example through compensatory techniques or through particular learning strategies). Whichever method is selected, theories of learning need to be consulted, understood and applied where necessary. In Baddeley's words, 'A theory of rehabilitation without a model of learning is a vehicle without an engine' (Baddeley, 1993, p. 235). It is now accepted that social, emotional, and cognitive functions are interlinked and difficult to separate (Wilson, Gracey, Evans & Bateman, 2009), so we should be aware of models of therapeutic change such as therapeutic working alliance, or the client's experience of being understood, which optimise learning and engagement in rehabilitation. Recent theories of identity (Gracey & Ownsworth, 2008), Compassion Focused Therapy (CFT; Ashworth, Gracey & Gilbert, 2011) and Narrative Therapy (White & Epston, 1990) are some of the latest additions to our treatment approaches.

As part of the complete process of rehabilitation, our treatments or interventions must be evaluated. Whyte (1997) pointed out that outcome should be congruent with the level of intervention. If intervening at the impairment (body structure and process) level then outcome measures should be measures of impairment and so forth. As most rehabilitation is concerned with the improvement of social participation, outcome measures should



reflect changes in this domain: for example, how well does someone who forgets to take medication now remember to take medication? We are continuously improving our evaluation of neuropsychological programmes but it is not easy. As Hart, Fann and Novack (2008) remind us, because of the great heterogeneity of patients receiving such rehabilitation and because of the variety of aims and methods required to achieve ultimate goals, the measurement of treatment effectiveness and final outcomes resulting from rehabilitation are difficult to evaluate (Hart *et al.*, 2008).

When considering the model, it is not difficult to recognise the labyrinth of theories to be negotiated in order to design treatment that is appropriate for each brain injured individual with his or her particular needs. The same is true when considering the circumstances and needs of that person's family and even in some cases the wider community. We have made considerable progress in recent times (Cicerone *et al.*, 2011) and are better at linking theory and practice (Wilson *et al.*, 2009) when designing practical solutions for the management of the brain injured person's daily life. These practical solutions are discussed in each chapter and supported by theoretical underpinning.

Despite considerable success, rehabilitation has to proceed with very limited resources. In some of the chapters, the survivors themselves comment on or allude to problems with funding. It is worth repeating once more that although rehabilitation for survivors of brain injury may seem to be expensive in the short term, it is cost effective in the long term. Given that most people who have survived an acquired brain injury and are referred for neuropsychological rehabilitation are young and will, on the whole, live a normal life span, they deserve to be given every chance to live as fulfilling a life as possible.

A major feature of the rehabilitation offered by the Oliver Zangwill team is its concentration on holistic treatment. A holistic approach to brain injury rehabilitation, 'consists of well integrated interventions that exceed in scope, as well as in kind, those highly specific and circumscribed interventions which are usually subsumed under the term "cognitive remediation"' (Ben-Yishay & Prigatano, 1990, p. 40). The holistic approach was pioneered by Diller (1976), Ben-Yishay (1978) and Prigatano (1986). Ben-Yishay and Prigatano (1990) provide a model of hierarchical stages in the holistic approach through which the patient must work in rehabilitation. These are, in order: *engagement*, *awareness*, *mastery*, *control*, *acceptance* and *identity*. The holistic approach argues that it is futile to separate the cognitive, social, emotional and functional aspects of brain injury. Given that how we feel emotionally affects how we think, remember, communicate and solve problems, and also influences how we behave, we need to acknowledge that these functions are inter-connected, often hard to separate and all need to be dealt with in rehabilitation. Holistic programmes offer both group and individual therapy to increase awareness, promote acceptance and understanding, provide cognitive remediation, develop compensatory skills and provide vocational counselling.

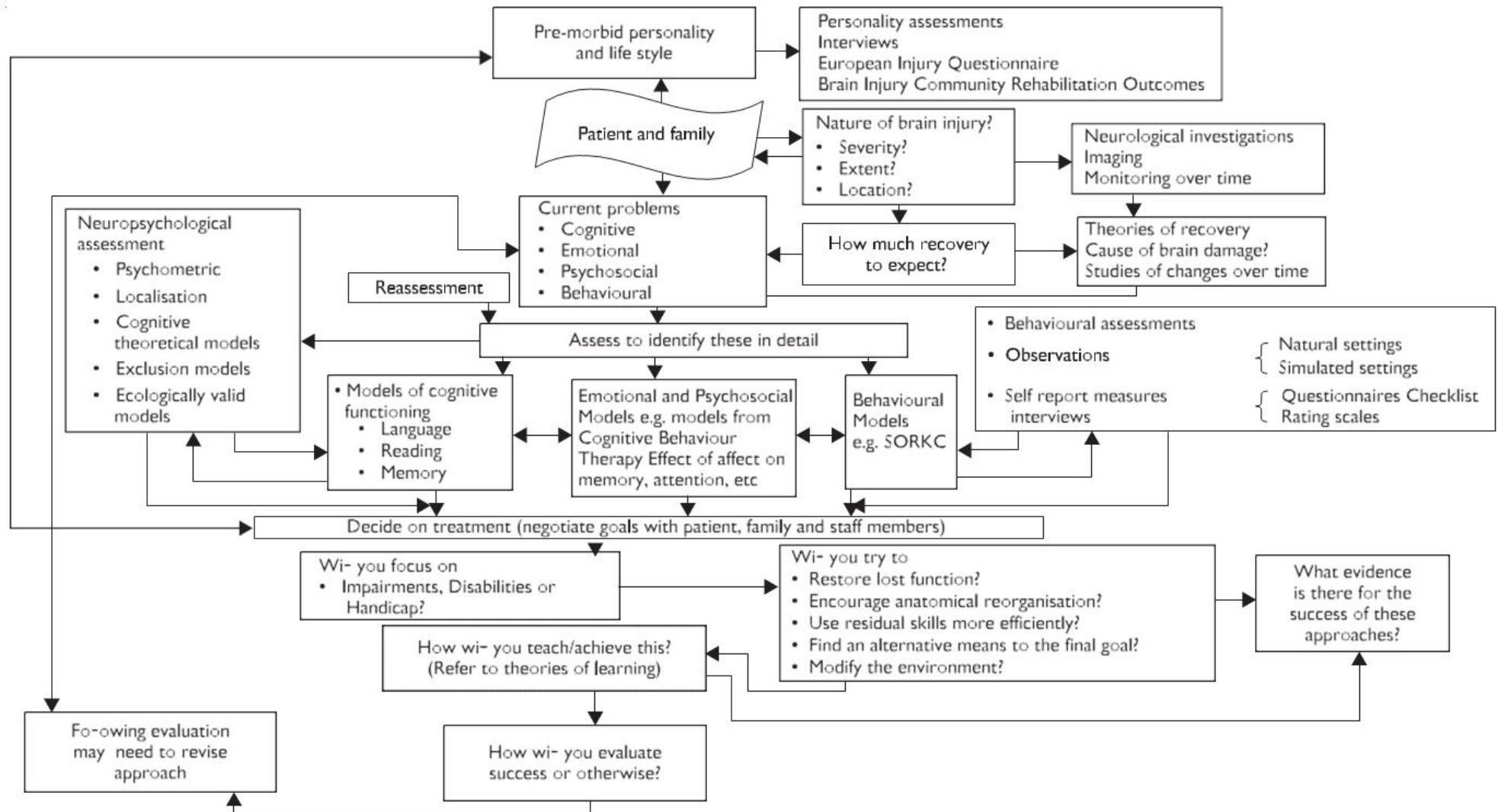


Figure 1.1 A model of Neuropsychological Rehabilitation (Wilson 2002, reproduced with permission of Taylor & Francis/Psychology Press).

Holistic programmes, explicitly or implicitly, tend to work through hierarchical stages, as described by Ben-Yishay and Prigatano (1990), and are concerned with:

- I. increasing the individual's awareness of what has happened to him or to her;
- II. increasing acceptance and understanding of what has happened;
- III. the provision of strategies or techniques to reduce cognitive problems;
- IV. the development of compensatory skills;
- V. the provision of vocational counselling.

All holistic programmes include both group and individual therapy. It can be argued that the holistic approach is less of a model and more of a series of beliefs, or, as Prigatano (1999) puts it, a series of 'Principles'. Nevertheless, it makes clinical sense and despite its apparent expense, in the long term it is probably cost-effective (Cope *et al.*, 1991; Mehlbye & Larsen, 1994; Wilson & Evans, 2002). In fact, there is mounting evidence that rehabilitation reduces the effect of cognitive, psychosocial and emotional problems, leading to greater independence on the part of the patient, reduction in family stress and eventual employability for many brain injured people (Cicerone *et al.*, 2005, 2011).

The programme at the Oliver Zangwill Centre (OZC) is based on these principles of holistic rehabilitation. Several chapters will refer to our assessment process and rehabilitation programme. Clients undergo assessment for programme suitability by attending the OZC for sessions with all team members that involve clinical interview, neuropsychological testing, mood assessment, family interview, community outings, functional tasks, participation in OZC meetings and lunch with current clients. Results are formulated as a poster that depicts the multiple contributions of personal and historic information, current context and the consequences of the injury on mood, cognition, communication and function.

The programme itself has evolved over time but essentially always includes attendance for 18 to 24 weeks with an intensive period devoted to psycho-education focused on themes including understanding brain injury, attention and memory, executive functions, communication and mood. Clients also learn and practise strategies dovetailed to their particular difficulties and receive individual psychological therapy to support them through the rehabilitation process. They then progress to the integration phase in which personal goals are developed and practised back in the client's own community. At present, clients attend the intensive phase for four days per week and the integration phase for two days per week, with follow up goal setting and reviews at three, six and twelve months post-discharge.

In their latest meta-analysis of neuropsychological rehabilitation, Cicerone and his colleagues (2011), as mentioned above, conclude that comprehensive holistic





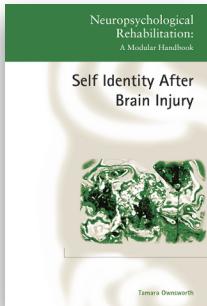
neuropsychological rehabilitation leads to improvement. They go even further in their guidelines when they say, 'Comprehensive-holistic neuropsychologic rehabilitation is recommended to improve post acute participation and quality of life after moderate or severe TBI' (Cicerone *et al.*, 2011, p. 526).

While this book is influenced by theory, it is the narrative of the survivors themselves that provides confirmation of the value of rehabilitation. White and Epston (1990) believe that stories or narratives shape a person's identity. Some of our survivors address this theme. We hope that their stories will help those working in rehabilitation to understand the principles involved in holistic brain injury rehabilitation and appreciate how these principles, combined with theoretical input, translate into clinical practice. In each chapter functionally relevant and meaningful goals, set by the therapist, the survivor and his or her family, are targeted; appropriate practical steps are taken; and outcomes evaluated. Finally, in acknowledging that rehabilitation is an interactive process between people with brain injury, health care staff and others, we give the survivors a chance to tell their own stories about their journey.



Overview of self-identity after brain injury

Chapter 2: Overview of self-identity after brain injury



The following is excerpted from *Self-Identity after Brain Injury* by Tamara Ownsworth. © 2014 Taylor & Francis Group. All rights reserved.

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This introductory chapter highlights the importance of addressing self-identity issues in brain injury rehabilitation and presents an overview of the book.

An injury to the brain can affect virtually any aspect of functioning. At the deepest level it can alter one's sense of self or the unique and persisting qualities that define who we are. People with brain injury may perceive losing some fundamental part of their selves (e.g., 'I'll never be a patch on who I was'). Close family members and friends may grieve the loss of the person and the relationship they once had. Changes to self after brain injury are complex and not readily observable or easy to address in rehabilitation. Neuropsychological rehabilitation broadly aims to assist people to manage the everyday consequences of brain injury and to live a meaningful and fulfilled life. All rehabilitation approaches contribute in some way to rebuilding a person's self-identity regardless of the aspect of functioning focused on (e.g., mobility, speech or memory). However, people do not reconstruct their sense of self from objective functional gains *per se*, but rather the *personal meaning* they derive from their everyday experiences after brain injury. The importance of self in rehabilitation has long been recognised, with neurorehabilitation and psychotherapy approaches used in combination to facilitate identity transition after brain injury (Ben-Yishay et al., 1985; Wilson et al., 2009; Ylvisaker, McPherson, Kayes & Pellet, 2008). Despite growing interest in self-identity after brain injury (Graeley & Ownsworth, 2008), there are few evidence-based guidelines to support identity-oriented assessment and intervention practices.

Research investigating changes to self in the context of brain injury has only emerged in the literature over the past few decades, stimulated largely by the seminal work on self-concept by Tyerman and Humphrey (1984). The interface between social psychology and cognitive neuroscience (i.e., social neuroscience) allows for a more advanced understanding of how sense of self emerges as a product of our neurobiology, culture and their interaction (Feinberg, 2011a; Jetten, Haslam & Haslam, 2012; Rochat, 2011; Walsh, Fortune, Gallagher & Muldoon, 2012). Neuropsychological models offer accounts of how cognitive and emotional subsystems of the brain work together to create an ongoing sense of self that actively constructs meaning in our day-to-day experiences (Conway & Pleydell-Pearce, 2000; Damasio, 1999; LeDoux, 2000).

In a keynote address at the Annual Brain Impairment Conference in Brisbane in 2010, Professor Barbara Wilson identified that research on self-identity after brain injury was one of the top ten cutting-edge developments in the field. In an article based on the address, she noted that: 'Contemporary models such as Conway's (2005) "self-memory

system" and Haslam et al.'s (2008) work on social identity theory provide a means of thinking about the interplay between brain systems, cognition, personal and social identity' (2011, p. 35). In light of such progress, it was timely to write the first book dedicated to self-identity issues after brain injury. This volume seeks to highlight recent developments in theory and research relevant to self-identity and consider the implications for clinical practice, thus providing a useful resource for students, clinicians and researchers in the field.

This book aims, firstly, to provide a context for understanding self-identity changes after brain injury by reviewing different theories of self and identity that derive from psychology, sociology and cognitive neuroscience. The second aim is to provide a comprehensive account of the impact of brain injury on self-identity. Thirdly, the book aims to review clinical strategies for assessing self-identity processes and interventions for supporting individuals to re-establish a positive identity after brain injury. The book's final aim is to summarise the main areas of progress in self-identity and brain injury research and identify directions to advance the field.

Overview of the book

Before considering the impact of brain injury on self-identity it is important to understand what is meant by 'self' and 'identity'. Chapter 2 provides an overview of historical perspectives and more contemporary theories that contribute to our current understanding of self-identity. This discussion draws initially on the ideas of early philosophers such as Aristotle and Locke, whose insights bear remarkable similarity to social neuroscience perspectives on self-identity today. The chapter then focuses on theoretical advances during the 19th and 20th centuries, including perspectives from psychoanalytic, developmental, humanistic and social psychology which collectively highlight the influence of biology, socialisation and culture. A discussion of more contemporary accounts of self and identity in the 21st century follows, based on cognitive and social neuroscience developments. In the final section of the chapter these perspectives are integrated into a framework which supports readers to consider the potential for changes to self after brain injury.

To introduce brain injury, Chapter 3 summarises the main causes and functional consequences and provides a developmental perspective. The diverse factors contributing to brain injury outcomes are conceptualised within a biopsychosocial framework, which recognises the interactive influence of pre-morbid, neurological, social environmental and psychological factors. The second half of this chapter focuses on the impact of sustaining a brain injury early in life and implications for emerging sense of self in childhood and adolescence. Empirical evidence regarding biological

and social vulnerability and factors moderating the relationship between the neuropathology of brain injury and functional outcomes is discussed. The psychosocial consequences of brain injury for children and adolescents are reviewed, with a specific focus on the impact of metacognitive and social cognition impairments on emerging sense of self. Two case studies (Dan and Jasmine) illustrate the complex interplay of factors influencing identity formation after childhood brain injury.

Chapter 4 presents a comprehensive review of psychological adjustment and self-identity changes after brain injury in adulthood. Psychological adjustment refers to the process of becoming aware of, making sense of, and adapting to changes in one's functioning and life circumstances. The inter-related processes of self-awareness, sense-making and coping are examined within a biopsychosocial framework. Building on this framework, research investigating self-identity changes after brain injury is specifically reviewed. Overall, this research indicates that premorbid characteristics and neuropsychological status influence perceived changes to self, and that opportunities to re-engage in meaningful activities and social roles are instrumental to rebuilding sense of self. An adaptive self-identity after brain injury is fostered by everyday experiences that provide personal meaning and reinforce self-worth.

Many different approaches are used in clinical practice to measure self-perceptions and other processes related to self-identity. The issues surrounding measurement of subjective phenomena are initially considered along with some caveats. Adopting the framework presented in Chapter 4, Chapter 5 initially reviews approaches for assessing self-awareness, sense-making appraisals and coping after brain injury. These approaches include self-report questionnaires, interviews and behavioural observation approaches. The focus then shifts to assessment of self-concept and changes in self-identity after brain injury, with an appraisal of methods developed specifically for the brain injury population. In the final section of Chapter 5, assessment approaches are considered for children and adolescents with brain injury. Approaches developed for the general paediatric population that have potential utility for brain injury are discussed with recognition that these methods require psychometric evaluation for this population.

The importance of focusing on identity issues in rehabilitation has long been recognised. Any intervention that aims to improve people's functioning and influence their self-perceptions contributes in some way to the identity reformation process. Chapter 6 reviews individual psychotherapy and neurorehabilitation approaches for adults with brain injury. Strategies for enhancing the working alliance and engagement in therapy are initially discussed. The application of and evidence base for diverse approaches, including psychoeducation and feedback, goal-directed interventions, cognitive and behavioural therapies and project-based learning are described. Further,



the utility of technological aids for supporting sense of self by enhancing attainment of goals and recall of everyday experiences is considered. Chapter 6 advocates for approaches that systematically integrate psychotherapy and neurorehabilitation to support emotional and cognitive functioning and participation in valued activities and relationships.

Chapter 7 emphasises the key influence of social factors and peer support in the identity reconstruction process, and provides a review of group, holistic and community-based interventions. The characteristics and efficacy of structured group interventions and comprehensive holistic rehabilitation programmes that create a therapeutic milieu are considered. A major challenge for clinicians is to support people with brain injury to maintain their gains after rehabilitation and build upon these outcomes through sustainable networks of support. Community initiatives that can provide a sense of belonging, achievement and contribution include paid work, volunteering, leisure activities, advocacy, group projects and leadership opportunities. Chapter 7 concludes with a discussion of the potential for social media to enhance social functioning and well-being of people with brain injury.

The effects of brain injury are far reaching, and can destabilise the entire family unit and the relationships and identity of its members. An essential aspect of rehabilitation involves supporting family members to adjust to the impact of the brain injury on their lives and well-being. The first section of Chapter 8 discusses the impact of brain injury on 'family identity' and reviews three main approaches to supporting family members. Approaches include: 1) involving family members in therapy for the person with brain injury, 2) interventions designed specifically for family members, and 3) family system interventions. The second part of Chapter 8 summarises rehabilitation approaches for children with brain injury. Despite a general absence of interventions that focus specifically on self-identity issues for children, related approaches in the literature include holistic neuropsychological rehabilitation, parenting or family-based interventions, and multi-component context-sensitive approaches (e.g., home, school and work).

The final chapter summarises and integrates leading developments in the self-identity and brain injury literature and provides directions for future research. Key advances in the field include: a) brain imaging studies mapping the neural correlates of self-related processing and social identification; b) theoretical accounts depicting the impact of brain injury on self-perceptions and identity; c) research revealing the role of biopsychosocial factors in identity reconstruction after brain injury; d) development and preliminary validation of measures assessing changes to sense of self; e) emerging evidence to support the efficacy of individual, group and community-based interventions for enhancing psychological adjustment to brain injury; and f)

family-based therapy and contextualised approaches for supporting people with brain injury and their family members. Priority areas for future research include: validating paediatric assessment tools, modelling the identity transition process over time and evaluating the efficacy of individual, group and family-based interventions for improving self-concept after brain injury. The chapter concludes the book by identifying future research directions along these lines.

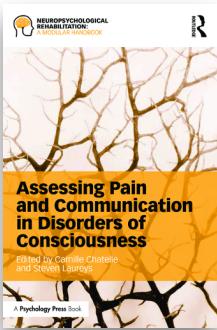
Conclusion

A brain injury is a life-altering experience that leads to changes in people's abilities and social situation. The most complex consequences entail changes in selfhood, which are perplexing for individuals, family members and professionals. Understanding and managing changes in self-identity is an integral part of rehabilitation. Although this has long been recognised, systematic research investigating changes to self in the context of neurological disorder has only emerged in the literature over the past two decades and such research is in its infancy. The focus on self and identity reformation therefore represents a relatively new frontier in brain injury research. It is hoped that this book stimulates research and clinical innovations that will enrich the lives of people with brain injury and their families.



Introduction to the challenge of pain and communication in disorders of consciousness

Chapter 3: Introduction to the challenge of pain and communication in disorders of consciousness



The following is excerpted from *Assessing Pain and Communication in Disorders of Consciousness* by Camille Chatelle and Steven Laureys. © 2016 Taylor & Francis Group. All rights reserved.

Camille Chatelle, Steven Laureys and Caroline Schnakers

Abbreviations

BCI	brain-computer interface
CRS-R	Coma Recovery Scale-Revised
DOC	disorders of consciousness
EEG	electroencephalography
MRI	functional magnetic resonance imagery
LIS	locked-in syndrome
MCS	minimally conscious state
MCS+	minimally conscious state plus
MCS-	minimally conscious state minus
PET	positron emission tomography
VS/UWS	vegetative state/unresponsive wakefulness syndrome

Learn more:



Advances in medicine for resuscitation and care have led to an increased number of patients surviving severe brain damage. Some of them will quickly recover consciousness and will be able to communicate, while others may evolve into various disorders of consciousness (DOC). In this introductory chapter, we will define the different DOC that can follow a severe brain injury and the clinical challenges associated with these patients in terms of pain management and detection of consciousness and communication.

Behavioral definition of DOC following a severe brain injury

Although no commonly shared definition of consciousness exists, it is widely accepted that it is a multicomponent term involving a series of cognitive processes such as attention and memory (Baars, Ramsey, & Laureys, 2003; Zeman, 2005). It has been suggested that consciousness is underlined by a large frontoparietal network, also called the global neuronal workspace (Baars, 2005; Dehaene & Changeux, 2011). Additionally, neuroimaging studies have highlighted the importance of the thalamo-cortical connections, especially in the emergence of consciousness (Tononi & Koch, 2008). Indeed, in patients who have recovered consciousness, a reestablishment of the correlation between associative cortices and the thalamus has been observed (Laureys et al., 2000b).

At the bedside, consciousness can be characterized by two main components: arousal and awareness (Laureys, Faymonville, & Maquet, 2002a; Posner, Saper, Schiff, & Plum, 2007). Clinically, arousal is manifested by spontaneous eye opening, whereas awareness is assessed by responses to external stimuli (e.g., command-following, visual pursuit, adequate emotional response). Although loss of arousal is associated with altered awareness (e.g., sleep, anesthesia), preserved arousal level does not necessarily imply preserved awareness. Following severe brain damage, a patient can either die (brain death) or evolve through different states of altered consciousness before possibly recovering full consciousness. Each of these states is associated with more or less severe cognitive and motor disabilities (see Figure 1.1). As shown in Figure 1.1, the restoration of spontaneous or elicited eye opening, in the absence of voluntary motor activity, marks the transition from coma to vegetative state/unresponsive wakefulness syndrome (VS/UWS). The passage from VS/UWS to the minimally conscious state minus (MCS-) is marked by the appearance of non-linguistic signs of conscious awareness. MCS plus (MCS+) patients show clear evidence of receptive or expressive language function. Emergence from MCS is signaled by the return of functional communication or object use. The locked-in syndrome (LIS) is the extreme example of intact cognition with nearly complete or complete motor deficit (Laureys, Perrin, Schnakers, Boly, & Majerus, 2005b). Each of these states has also been studied in terms of brain metabolism at rest or brain activation in response to external stimuli.

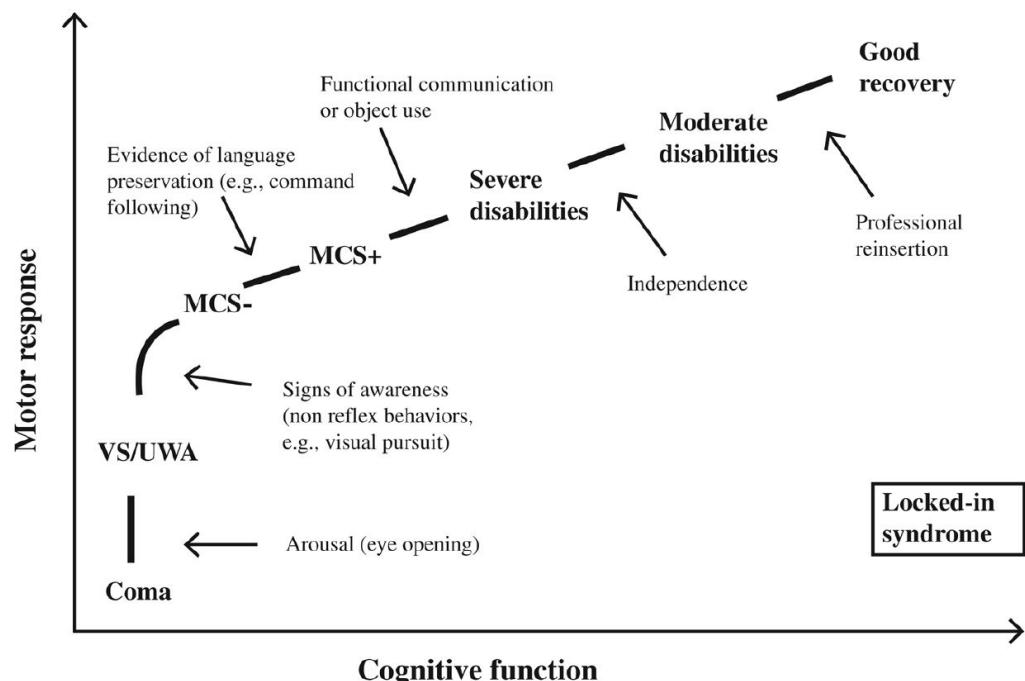


Figure 1.1 Different clinical entities encountered on the gradual recovery from coma, illustrated as a function of cognitive and motor ability (adapted from Chatelle & Laureys, 2011).

Brain death

The term “brain death” suggests that the organism cannot function as a whole. Critical functions such as respiration, blood pressure, neuroendocrine and homeostatic regulation, and consciousness are permanently lacking. The patient is apneic and unreactive to environmental stimulation (Guidelines for the determination of death, 1981). This term can only be used after bedside demonstration of irreversible cessation of functions of the brain and the brainstem. Brain death is usually caused by a severe brain lesion (e.g., massive traumatic injury, intracranial hemorrhage, or anoxia) resulting in an intracranial pressure higher than the mean arterial blood pressure. The diagnosis can be made within 6–24 hours, after excluding pharmacological or toxic treatments or hypothermia as potential confounders. This state is characterized by the absence of residual brain metabolism, confirming the absence of neuronal function in the whole brain (Laureys, Owen, & Schiff, 2004a).

Coma

Some patients can remain in a coma, neither aroused nor aware, for several weeks. Their eyes are constantly closed and they do not manifest voluntary behavioral responses. Generally, patients emerge from their comatose state within two to four weeks (Posner et al., 2007). The prognosis is influenced by different factors such as etiology, the patient’s general medical condition, and age. Outcome is most likely to be bad if, after three days of observation, there are still no pupillary or corneal reflexes, there is stereotyped or no motor responses to noxious stimulation, and an isoelectrical or burst-suppression electrophysiological (EEG) pattern is observed. Prognosis in traumatic coma survivors is better than in anoxic cases (Whyte et al., 2009). Recovery from coma may lead to a vegetative state, a minimally conscious state or, more rarely, to a locked-in syndrome (Bruno, Vanhaudenhuyse, Thibaut, Moonen, & Laureys, 2011; Posner et al., 2007). Coma is generally associated with a global decrease in brain metabolism of 50–70% of the normal range.

Vegetative state/unresponsive wakefulness syndrome

The “vegetative state” (VS), “an organic body capable of growth and development but devoid of sensation and thought,” was defined in 1972 by Jennett and Plum (Jennett & Plum, 1972). The term was proposed to describe a state in which autonomic functions (e.g., cardio-vascular regulation, thermoregulation) and arousal (wakefulness and rest cycles) were preserved with the absence of awareness. Behaviorally, patients in VS



open their eyes spontaneously or in response to stimulation, but they only show reflexive (involuntary) responses to the environment. Recently, some have suggested replacing the term VS with “unresponsive wakefulness syndrome” (UWS) in order to avoid the negative association with the word “vegetative” and to better describe the behavioral pattern observed in this population (Laureys et al., 2010). It has been suggested that the state can be defined as permanent when there is no recovery after a specified period (three or twelve months, depending on etiology, anoxic or traumatic, respectively) (American Congress of Rehabilitation Medicine, 1995; Jennett, 2005). However, further evidence suggests that patients in a VS/UWS can recover even after this length of time, so the term may therefore not be appropriate (Estraneo et al., 2013; Estraneo, Moretta, Loreto, Santoro, & Trojano, 2014). Studies on global brain metabolism have usually shown a decrease of about 40–50% of normal range values in VS/UWS patients (Stender et al., 2015), although some studies have reported cerebral metabolism (Schiff et al., 2002) or blood flow (Agardh, Rosen, & Ryding, 1983) in the normal range in some cases.

Minimally conscious state

Patients in a minimally conscious state (MCS) are aroused and manifest fluctuating but consistent and reproducible signs of awareness (Giacino et al., 2002). Visual pursuit appears to be an early behavioral marker of the transition from VS/UWS to MCS (Giacino & Whyte, 2005), but these patients can also show other voluntary behavioral responses and/or oriented emotional reactions such as responses to verbal orders, object manipulation, oriented responses to noxious stimulation, visual fixation, or appropriate crying/smiling. However, responsiveness can fluctuate, which makes the detection of voluntary behaviors at the bedside challenging. Some researchers have recently proposed subcategorizing MCS into MCS minus (MCS-) and MCS plus (MCS+), based on evidence of differences in brain metabolism within the language network (Bruno et al., 2012; Bruno et al., 2011). MCS+ would encompass patients who show clear evidence of receptive or expressive language function (e.g., command-following). In contrast, MCS- would define those who demonstrate only nonlinguistic signs of conscious awareness (e.g., visual pursuit). The specific behaviors required to meet the criteria for MCS+ and MCS- are still being discussed and additional empirical investigation is needed before these categories can be implemented in clinical practice.

Emergence from MCS is defined by the recovery of functional communication and/or functional use of objects (Giacino et al., 2002). Some patients can remain in MCS without fully recovering consciousness for a prolonged period (Fins, Schiff, & Foley, 2007). Prognosis for recovery in MCS patients remains very difficult because of the marked heterogeneity in the underlying pathophysiology of these patients. However, a

better outcome for MCS as compared to VS/UWS patients has been reported (Lammi, Smith, Tate, & Taylor, 2005; Luauté et al., 2010). If the global metabolic rate remains usually higher in MCS than in VS/UWS (Stender et al., 2015), it does not always show substantial changes and return to normal after recovery of consciousness (Laureys, Lemaire, Maquet, Phillips, & Franck, 1999). However, activation studies reported differences between MCS and VS/UWS patients, suggesting a disparity between cognitive preservation and processing of external stimuli such as pain (see also Chapter 2 and 3).

Locked-in syndrome

Not to be misidentified as patients suffering from an altered state of consciousness, locked-in (LIS) patients cannot move or talk but they are usually able to use vertical eye movements and/or blink to communicate. This syndrome is often due to a selective supranuclear motor deafferentation that results in the paralysis of all four limbs and the last cranial nerves without interfering with consciousness (American Congress of Rehabilitation Medicine, 1995; Plum & Posner, 1983) or cognition (Schnakers et al., 2008b). Therefore, these patients may present the same behavioral pattern as that observed in VS/UWS or MCS patients, which often leads to misdiagnosis of altered state of consciousness (see also Chapter 8). LIS can be subcategorized based on the extent of motor impairment (Bauer, Gerstenbrand, & Rumpl, 1979): patients with classical LIS are totally immobile but can make vertical eye movements and blink; incomplete LIS is characterized by remnant nonocular voluntary motions (e.g., head or finger movements); and total LIS patients are completely paralyzed and have no eye movement (see also Chapter 6). When looking at the brain metabolism of these patients, researchers observed no significant decrease in metabolism in the supratentorial gray matter as compared with healthy subjects (Laureys et al., 2005a). However, a positron emission tomography (PET) study by Levy et al. reported a 25% reduction in cerebral metabolism in three LIS patients as compared to healthy controls (Levy et al., 1987).

Cortical processing in disorders of consciousness

Several neuroimaging studies using PET and functional magnetic resonance imagery (fMRI) have studied brain processing in response to stimulation in patients with DOC. Following auditory and nociceptive stimulation, limited brain activation was reported in a majority of VS/UWS patients, whereas MCS patients showed an activity level close to that observed in control subjects (Boly et al., 2005; Boly et al., 2008; Laureys et al., 2000a). More specifically, activation studies performed on a VS/UWS group using auditory stimulation (i.e., tones) showed preserved functioning of the primary auditory

cortex but without involvement of other brain areas (such as the temporo-parietal junction; Laureys et al., 2000a). Following noxious stimulation (i.e., electrical stimulation of the median nerve), an increase in activity could be observed in the midbrain, contralateral thalamus, and primary somatosensory cortex, but again with limited activation in higher-order brain areas more deeply involved in perception processing (Kassubek et al., 2003; Kotchoubey et al., 2013; Laureys et al., 2002b; Markl et al., 2013; see also Chapter 2). Additionally, low-order primary cortical activity seemed to be isolated from higher-order associative cortical activity in VS/UWS patients (Laureys et al., 2002b). On the other hand, auditory and nociceptive stimulations of MCS patients led to widespread activation involving associative cortices considered hierarchically superior in the processing of sensory information. Such activation suggests that these patients have partially preserved auditory and pain processing (Boly et al., 2005; Boly et al., 2008; Laureys et al., 2000a; Schiff et al., 2005). Other studies have also reported wide-spread activation in the temporal, insular, and amygdala areas in response to stimuli with emotional valence (Bekinschtein et al., 2004; Laureys et al., 2004b).

Clinical diagnosis: pain and communication

While neuroimaging studies offer a great means to better understand brain processing in DOC, the majority of the findings is based on group analyses and can be difficult to apply to individual patients in clinical settings. Additionally, it remains difficult to interpret the significance of partially or fully preserved brain activation in terms of a patient's conscious awareness of the self and the environment and his or her subjective experience.

In terms of pain management in patients with DOC, the assessment is limited by the difficulty or impossibility of establishing functional communication. The inability of the patient to provide a subjective report leads to potential ethical and medical concerns. In both acute and chronic stages, several conditions – such as polytraumatic injuries, open wounds, and spasticity – are likely to induce pain, especially during care and mobilization (Chatelle et al., 2014). The need for analgesic treatment and close monitoring is therefore evident, and especially in light of previous studies suggesting pain perception capacity in patients with DOC. A European survey asked over 2,000 medical and paramedical professionals about their beliefs regarding possible pain perception in patients with DOC. Their answers regarding whether or not these patients (VS/UWS or MCS) can feel pain were very different. Interestingly, the opinions of these healthcare professionals varied according to their profession, religious beliefs, and region of origin (Demertzi et al., 2009). The disparity in opinions and perceptions of pain among caregivers may lead to variability in pain and symptom management. Given



the challenges of defining levels of consciousness in this population (Schnakers et al., 2009) and considering the degree of clinical uncertainty regarding pain perception, pain treatment should be considered for all patients (Chatelle et al., 2012; Monti et al., 2010; Schnakers et al., 2009) and should be managed using standardized procedures (i.e., behavioral scales) (Chatelle et al., 2014; see also Chapters 3 and 4). However, until now no clear guidelines for pain management in cases of DOC have been proposed.

In the context of pain assessment, obtaining an accurate diagnosis of the patient's level of consciousness and communication is really important. The patient's quality of life and outcome may depend on an accurate assessment at the bedside. In addition to differences in brain processing in VS/UWS and MCS patients, outcome studies have reported that MCS patients usually have a better prognosis than those with VS/UWS (Katz, Polyak, Coughlan, Nichols, & Roche, 2009; Luauté et al., 2010). In addition, some patients might be diagnosed as VS/ UWS or MCS despite their suffering from LIS (see Chapter 6) and therefore processing fully painful conditions.

Currently, the clinical diagnosis of consciousness is mainly based on behavioral assessment at the bedside. The detection of oriented/voluntary responses and functional communication is really important for detecting consciousness as both signs of response and communication, respectively, indicate emergence from VS/UWS and MCS. Functional communication also differentiates LIS from VS/UWS patients. However, the difficulty of objectively distinguishing reflexive from voluntary responses makes the assessment very challenging for clinicians.

In this context, the use of standardized tools can help the examiner to limit errors associated with subjectivity (see Chapter 3). Schnakers et al. (2009) reported that 41% of patients were diagnosed incorrectly by expert team consensus, when compared to the diagnoses obtained using a standardized assessment instrument, the Coma Recovery Scale-Revised (CRS-R) (Giacino, Kalmar, & Whyte, 2004). This data suggests that the use of a sensitive scale is crucial when assessing consciousness (Schnakers et al., 2008a).

On the other hand, standardized behavioral assessments are complicated by motor disabilities, aphasia (Majerus, Bruno, Schnakers, Giacino, & Laureys, 2009), fluctuation in arousal level and vigilance (Giacino et al., 2002), and other physical impairments such as blindness or deafness. Because of these compromising factors, it can be very difficult to make an accurate diagnosis. It is therefore important to develop other paraclinical tools to detect signs of consciousness and communication when no response can be observed at the bedside. As misdiagnosis can lead to grave ethical consequences, especially in terms of end-of-life decision-making (Andrews, 2004) and pain treatment (Chatelle et al., 2014), additional tools should be used to assess remnant cognitive abilities in patients with DOC.

In this context, new technologies such as electroencephalography, PET, or fMRI could provide paraclinical tools that bypass the motor pathway and allow clinicians to detect consciousness. In the last decade, studies have reported the potential of these techniques to probe command-following in severely brain injured patients by asking them to actively modulate their brain activity during a specific task (Chatelle et al., 2012; Chatelle, Lesenfants, Guller, Laureys, & Noirhomme, 2015). By extension, these methods could be used to control a brain-computer interface (BCI). BCI, by definition, uses brain activity alone to drive external devices or computer interfaces (Wolpaw, Birbaumer, McFarland, Pfurtscheller, & Vaughan, 2002). Recent studies have shown the usefulness of BCIs in controlling motor prostheses, cursors, access to the Internet, and communication (Citi, Poli, Cinel, & Sepulveda, 2008; Lee, Ryu, Jolesz, Cho, & Yoo, 2009; Mugler, Ruf, Halder, Bensch, & Kler, 2010; Müller-Putz & Pfurtscheller, 2008; Sellers, Vaughan, & Wolpaw, 2010; Yoo et al., 2004). In this context, the development of accurate BCIs enabling command-following or even a simple binary communication code would have a huge impact, from a clinical perspective as well as from a social one (see Chapter 5). BCI may make it possible for some behaviorally noncommunicative patients to participate in their treatment, to tell their caregivers/family how they feel and when/where they are in pain, and also to report about their desire to live or to die. However, the introduction of these technologies in clinical settings will also raise important ethical discussions (see Chapter 7).

In this book, we aim to provide clinicians with the most up-to-date information in the field of clinical diagnosis of pain and communication in DOC, including ethical reflections and evidence-based recommendations for managing care in this population.

Notes

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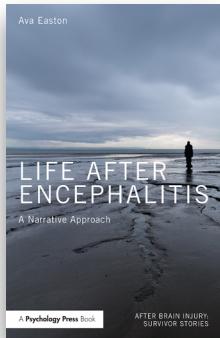
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Encephalitis: what it is and what it does

Chapter 4: Encephalitis: what it is and what it does



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I was discharged home and had to believe that my husband was the right person, but I had no sense of certainty about him. Although I could not recognize our children I did have the belief that they were mine. I have memories of that time which are very frightening and upsetting. I had no sense of belonging to this family who I was told was mine. I remember feeling very frightened about the identity of my husband in particular and very separate from a unit of individuals who seemed so intense and confident together.

Encephalitis survivor: Rytina, 2007: pp. 18–19

Introduction

Encephalitis is a thief. In the same way that we have watched in recent months Ebola rob people of their lives and those they care about, encephalitis has quietly been at work for hundreds of years, robbing families of their loved ones, and even in those families where the person survives, it robs them of the person they once knew. Encephalitis steals survivors' capacity to remember as well as their personalities and the types of abilities we generally take for granted: memory, concentration, attention, thinking, judgement, inhibition. For many there are additional outcomes such as epilepsy and levels of fatigue so great that returning to work or education will remain elusive. This is, of course, where the person survives; many don't.

This chapter describes what encephalitis is; considers its global presence; and the outcomes for survivors and their families. We look in brief at diagnosis, treatment and management of the condition. In particular this chapter considers the psychosocial implications for people affected and issues around quality of life. As this chapter draws to a conclusion we will have begun to develop an understanding of encephalitis and why people's narratives may be important. In order to give this a broader context Chapter 3 will look in more detail at narratives and their role in medicine more generally.

What is encephalitis?

Encephalitis is a complex illness in its presentation, diagnosis, prognosis and long-term consequences (Solomon *et al.*, 2012). Encephalitis is inflammation of the brain tissue and is a syndrome with hundreds of potential causes. It can occur at any age, in any part of the world and is caused either by infection, usually viral, or by autoimmune disease. It is useful to think about encephalitis as having two primary causes: infection and autoimmune malfunction. There are two primary sub-causes of autoimmune encephalitis (post-infectious and other non-post-infectious causes) and finally there is a fourth type where people are progressively ill and sadly, in many cases, die. These

descriptions can be a little more understandable if you think of them as four sub-sets (see Table 2.1).

Table 2.1 Encephalitis sub-sets

<i>Infectious encephalitis</i>	<i>Post-infectious (autoimmune) encephalitis</i>
This type of encephalitis is caused when someone has an infection which breaches the blood-brain barrier, mounting a direct infectious attack on the brain tissue.	This type of encephalitis is caused when someone has had an infection or vaccine and their own immune system overreacts and launches an attack on the tissue of the brain.
<i>Autoimmune encephalitis</i>	<i>Sub-acute, chronic and slow</i>
This type of encephalitis is caused when a person's own immune system overreacts to something in the body it considers is alien. For example in response to a tumour or anti-neuronal antibodies.	These types of encephalitis are often terminal and can result for a variety of reasons. Examples include being unable to identify the cause of an autoimmune reaction or as the result of a persistent infection (for example a mutated measles virus resulting in sub-acute sclerosing pan-encephalitis (SSPE)).

Damage to the brain is caused by the direct protagonist (for example an infection) and also by the resultant inflammation and swelling of the brain.

Infectious and autoimmune encephalitides can present in quite dramatically different ways. Table 2.2 illustrates the symptoms that might present in the two different types.

Table 2.2 Symptoms of encephalitis

<i>Infectious encephalitis</i>	<i>Autoimmune encephalitis</i>
<p>Infectious encephalitis often has a rapid onset.</p> <p>Flu-like symptoms</p> <p>Dizziness</p> <p>Malaise</p> <p>Headache</p> <p>Vomiting/gastro-intestinal upset</p> <p>Fever</p> <p>Later stages indicating a more serious illness involve lowered consciousness which may include:</p> <p>Confusion/Drowsiness/ Seizures/Coma</p> <p>Other symptoms may include:</p> <p>Photosensitivity/Sensory Change/ Inability to speak or control movement Uncharacteristic Behaviour</p>	<p>Autoimmune types of encephalitis can have a longer onset than infectious causes.</p> <p>Symptoms will vary depending on the cause but may include:</p> <p>Confusion Altered personality or behaviour Psychosis Movement disorders Repetitive, involuntary motor or vocal tics Seizures Hallucinations Memory loss Sleep disturbance</p>

If you begin to think about all the sources of infection that exist (viruses, bacteria, parasites, fungi) the complexity and extent of this condition, and the problems it poses for diagnosis and treatment begin to become clear.

The numbers

For many years statistics around incidence and prevalence were scarce to non-existent. Global incidence figures are difficult to ascertain due to variations such as geographic distribution of causative agents, immunisation policies of different countries and methodological issues such as how cases are defined, diagnosed and recorded. A paper by Jmor *et al.* (2008) is probably the best attempt at global incidence figures: a minimum of 10 per 100,000 in children, 2 per 100,000 in adults and a minimum incidence of 6 per 100,000 for all age groups. If we assume a global population of 7.125 billion (www.google.co.uk/publicdata/explore?ds=d5bnc_ppjof_8f_9_&met_y=sp_



[pop_totl&hl=en&dl=en](#), accessed 9 July 2015) then we can estimate a minimum of 4.25 million cases of encephalitis (for all age groups) a year globally.

In 2013 the first realistic figures for incidence were released for England. A study conducted by Public Health England estimates (based on capture–recapture models) an incidence of 5.23 cases/100,000/year, although the authors state the models' indicated incidence could be as high as 8.66 cases/100,000/year (Granerod *et al.*, 2013). This means there are likely to be in the region of 6000 people diagnosed with encephalitis in the United Kingdom each year. That's 16 people every day. This, it is suggested, is also considered an underestimate. Not only that but the study suggests (again considering it an underestimate), that encephalitis is costing the NHS around £40 million a year. This figure does not include the costs of rehabilitation, long-term care and the loss to the economy from those of working-age unable to return to work (Granerod *et al.*, 2013).

This means that encephalitis has a higher incidence than motor neurone disease (Hoppitt *et al.*, 2011) and certain forms of meningitis ([www.meningitis.org/facts](#), accessed 9 July 2015). Yet, despite these conditions being less common, they receive a much higher clinical and public profile than encephalitis. People have often not heard of encephalitis unless it has happened to them or they are caring for a survivor.

Mortality is higher than many other neurological diseases (Chaudhuri and Kennedy, 2002; Raschilas *et al.*, 2002), and may be even higher than necessary in resource-poor countries which lack vaccination programmes for preventable forms (see section below on prevention). Rates of recovery following encephalitis for those who survive are less than for some other forms of acquired brain injury (Moorthi *et al.*, 1999; Pewter *et al.*, 2007). The consequences of encephalitis and its impact upon the people it affects (directly and indirectly) are more often than not social in nature and poorly understood (Easton *et al.*, 2006; Stapley *et al.*, 2008). What follows is a brief and broad account of these consequences, providing context in which the use of narratives of those affected by encephalitis can be explored in more detail in the chapters that follow.

Diagnosis

Diagnosis includes a number of tests and is often considered a diagnosis of exclusion (other conditions are ruled out one by one). Generally a full patient history will be important: what is their health status (do they have any conditions that might affect their immunity – for example HIV; do they have a history of mental health problems, drug or alcohol use?); have they been exposed to any infections recently (for example

measles, mumps or chickenpox)?; have they been abroad recently (for example could they have been bitten by ticks or mosquitoes that may have been carrying infection)?; what is their occupation (for example have they been exposed to any toxins or chemicals)? As a matter of course, blood will be taken and patients are (or should be) tested for HIV (Nightingale *et al.*, 2013). Patients will often be sent for brain imaging (CT or MRI) to ascertain if there is any inflammation of the brain and its extent. Electro-encephalograms (EEG) are used to establish patterns of brain activity. Finally, and perhaps crucially, a lumbar puncture (or spinal tap) will be conducted where it is safe to do so. This involves inserting a needle in the base of the spine and extracting some of the fluid that bathes the brain and spinal cord (cerebro-spinal fluid). This can then be tested to identify if infection is, or has been, present. Some types of autoimmune encephalitis can be diagnosed through specific tests measuring the presence of antibodies (for example anti-NMDA receptor encephalitis) or through identifying the presence of a tumour. In some cases where diagnosis is proving more difficult, a biopsy of some tissue of the brain will be performed in order to conduct further testing in an attempt to identify the cause. There is no single diagnostic test for encephalitis so all of these methods are used to help create a picture that will contribute toward diagnosis.

Researchers across the world are working hard to develop methods to better diagnose encephalitis in all its various forms and excellent progress has, and continues to be made, toward producing guidelines (Britton *et al.*, 2015; Kneen *et al.*, 2012; Solomon *et al.*, 2012). See also the biographies and narratives of Professor Tom Solomon and Dr Sarosh Irani in Chapter 4, for details of some of the latest and ongoing work.

Treatment

Treatment will vary dependent upon the cause. Where infectious types of encephalitis are suspected a drug called Aciclovir is administered. This is really only useful where the herpes virus is the cause (the herpes simplex virus which causes cold sores is a key protagonist in encephalitis); however, it is generally given as a precaution as soon as encephalitis is suspected. Other viral infectious causes generally have to run their course and depend on the person's own immune system being strong. Other infectious causes such as bacterial or fungal are much rarer but would be treated with antibiotics or anti-fungal treatments as appropriate. In autoimmune types patients are treated with a range of immune-modulatory therapies such as high-dose steroids, immunoglobulin and plasma exchange. Where there is an underlying cause such as a tumour then it will need to be removed or treated to help stop the autoimmune response.

Outside of this, treatment is symptomatic and usually involves the type of nursing care used in very poorly patients (for example ventilation, monitoring of consciousness and respiration, sedation, anti-convulsants and treatments to address secondary infections along with keeping the patient hydrated).

Professor Solomon and Dr Irani discuss in more detail the diagnosis and treatment of infectious and autoimmune types of encephalitis (see Chapter 4).

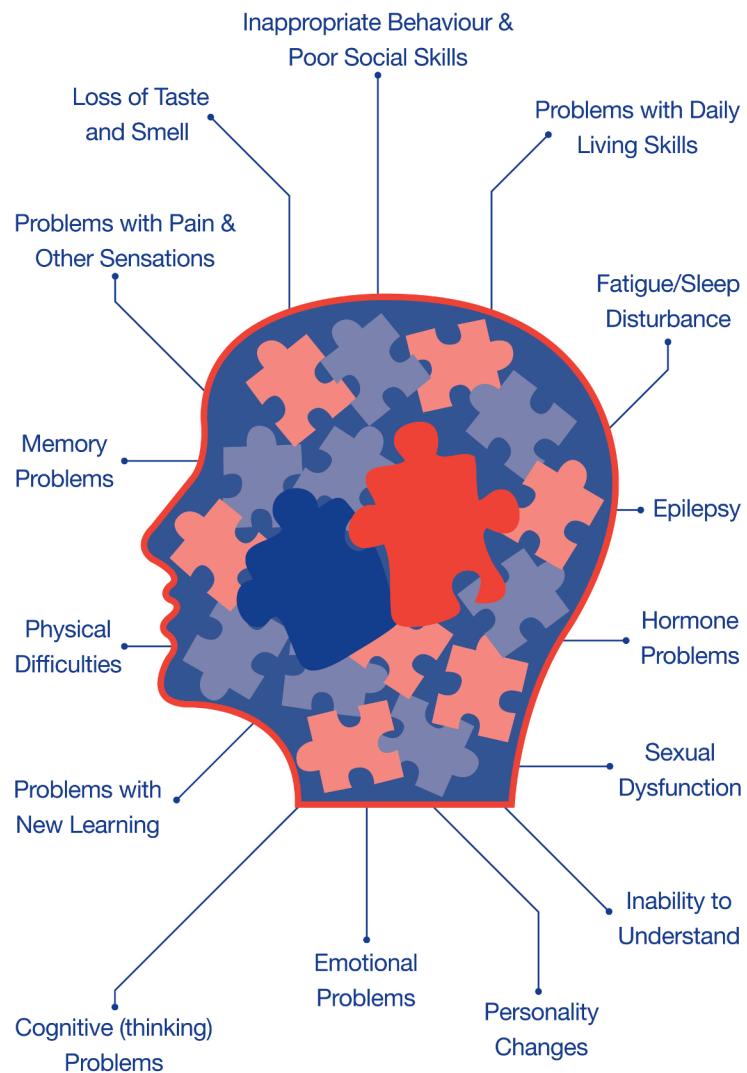
Prevention

Some forms of encephalitis are preventable. For example tick-borne encephalitis and Japanese encephalitis are vaccine-preventable. Travellers, in particular those from many Western countries, have access to vaccinations via their health services or travel clinics, if they are travelling to areas where these types of encephalitis may be endemic, particularly rural areas. Despite this an increasing number of people are travelling to what they consider ‘harmless’ locations such as Austria, Sweden and Switzerland for walking holidays without considering vaccination against tick borne encephalitis (www.encephalitis.info/files/8813/8694/8907/Tick-borne_Diseases_on_the_Map.pdf, accessed 9 July 2015). Vaccination programmes can dramatically drive down incidence in endemic countries. However, in resource-poor countries where, for example, Japanese encephalitis is prevalent, delivering vaccination programmes to indigenous populations can prove challenging (Michael and Solomon, 2012). In addition many environmental prevention measures can be taken such as using sprays to deter vectors (ticks and mosquitoes), using mosquito nets and full clothing protection, and in other instances such as rabies encephalitis, avoiding engagement with animals such as dogs and bats.

The consequences of encephalitis

Whilst some people may make a good recovery, the long-term consequence of encephalitis for many is injury to the brain. Brain injury may manifest in a number of ways, in particular cognitive, physical, behavioural, emotional and psychosocial problems are all umbrella terms under which a vast array of difficulties may occur (Williams and Evans, 2003; Wilson *et al.*, 2015). These are illustrated in Figure 2.1 below. As well as issues with fatigue and epilepsy, people who are affected by encephalitis may experience changes in their view of themselves (or their relatives): they are not the person they were before (Atkin *et al.*, 2010; Segal, 2010). Memory problems may result in a lack of continuity and order to their lives, in particular for those with loss of memory. Changes in personality, a sense of feeling different, as well as loss of control over emotions, thoughts or actions, can be upsetting. People may





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Figure 2.1 The consequence of encephalitis (reprinted with kind permission: The Encephalitis Society).

Brain injury: a hidden disability

If a person presents with a broken leg, everyone can see it as well as the related medical interventions and treatments, and people are also, over time, able to witness progress and recovery (Ponsford and Fleminger, 2005; Stone, 2005). Understandably, during the acute illness, physicians are focused upon saving life and limiting damage. The patient is usually too ill at this point to be capable of, or interested in, engaging in any meaningful way with their treatment or medical team (Rier, 2000). However, once the acute phase of encephalitis has subsided then the time following this is an important but often neglected feature of encephalitis. People are often discharged from hospital once they appear to have made functional progress: for example they can walk and talk. The possibility of acquired brain injury being a consequence of encephalitis is often not discussed, and so when people return to their communities and find they are no longer able to do everyday tasks and participate in their usual social relationships and activities, they have little understanding of what they are coping with and are left frightened and confused.

The primary brain injury can also be combined with secondary psychological difficulties, and Webb (1998) suggests the lack of an obvious physical impairment is “poor compensation for the sequestration of the mind” (p. 543), suggesting “those who are mind-impaired are consigned to the wastelands of social exclusion” (p. 549). A gradual and painful process of adjustment may follow, when the person realises the full extent of their residual disabilities and an appreciation that they may be unable to recover former skills and lifestyle. This process of recognition may result in periods of anger, grief and depression, characterised by mood swings, confusion, frustration and uncertainty. Such reactions are delayed for some people who understandably find it hard to accept their limitations. They may respond with a renewed determination to make a full recovery, and may set themselves unrealistic targets, leading to repeated disappointment and despair (Easton et al., 2006).

Complicating people’s experiences further, there is commonly an oft-quoted ‘story’ of how a person’s brain injury occurred, for example a road traffic accident, a sporting injury or as the result of an assault. This is not so for encephalitis. In addition in these early days of recovery the person affected may have no memory of their illness onset and families are often confused and lacking in the information required to understand and recount what has happened to their loved one. This results in a degree of uncertainty that shakes people’s foundations, and the impact and devastation people experience when they can no longer rely upon their own minds or memories should not be underestimated. This lack of an event or accident, along with no memory of their illness onset, is significant and results in a lack of a ‘story’. Understandably,



descriptions of brains and infections are not attractive, and can be frightening for audiences, particularly where fears around contamination or contagion might exist. Resultantly people have fears not only about the future but also about the responses of other people if they explain what has happened to them. Encephalitis strikes at the core of who people are, creating uncertainties, self-doubt and biographical disruption (Atkin et al., 2010). This, in turn, along with the hidden nature of brain injury all has an impact upon people's social experience and their identity, a concept considered in this next section.

Identity and social consequences

Illness is, as I have already suggested, negotiated within a wide social context that includes relationships, family dynamics, the wider community and environment. Our identity is mixed up within a social context – identity is not just about who we think we are, but also who others think we are, and who we think others think we are (Jenkins, 2006). Our identity then is negotiated within a social context: people have to make sense of what is happening to them in relation to others. Their illness experience therefore is socially negotiated. Particularly in encephalitis it is important to remember that the condition is variable both in its clinical consequences and also in relation to how those affected make sense of their condition (Stapley et al., 2008). In other words, some people are severely affected by the condition and this is reflected in how they describe themselves and the outcomes they experience, while some people report little impact. In addition to this, a person may cope well and distance themselves from the significance of the illness, while at other times they are the illness. These, however, are two separate processes, which will often change over time. The array of problems following encephalitis may be responsible in some cases for people withdrawing from their usual networks and communities, due either to changes in mood and behaviour or because people no longer trust themselves or their abilities when interacting with others (Williams and Evans, 2003). It is also of course important to consider the impact upon the family. Brain injury is a very direct and personal thing, however, it is also interpreted by, and has consequences for, the family and often the wider community (Lezak, 1988). As well as difficulties with memory, mood and behaviour, changes in social interaction may also be seen (Dewar and Gracey, 2007). Therefore people may experience less understanding from family, friends, professionals and the wider society leaving them to feel disrespected and powerless (Lorenz, 2010).

The family of a person with encephalitis often struggle to offer them support, while at the same time coming to terms with changes in their loved one. Family members and carers provide practical, social and emotional support and this can have important implications for their own lives. They are doing things for a relative which they were



not doing before. This can include anything from personal care to assuming moral responsibility for another person. Research by Man (2002: p. 1026) on family caregivers' reactions and coping, confirmed that "families' psychological reactions, such as depression and irritability, have been reported to be intense, particularly in relation to the presence of behavioural problems and physical impairment in the injured individual". Ponsford et al. (2003) established that anxiety and depression were more likely to be present in those responsible for the care of a brain-injured relative. Webb (1998) observed what he termed the "bankruptcy of identity" (p. 550) – a loss of sense of self, or a loss of self-identity that family members often bear witness to in respect of their brain injured family member. A family member's illness can mean a carer might also have to renegotiate their own sense of identity. This may include re-evaluating values and assumptions associated with their relationships (that they previously took for granted). This means a person has to reconstruct some sense of purpose or meaning within their life, all complicated and at the same time realised within the context of their personal and social relationships. Many individuals and their family members do make progress in recovering and subsequently understanding what has happened to them. This is, however, often not done in isolation but with the support and help of those around them (Haslam et al., 2009). In this next section we consider outcomes and quality of life post-encephalitis in more detail.

Outcomes and quality of life

There is a dearth of literature on the longer-term implications of encephalitis generally; however, a small but growing literature is beginning to establish that the outcomes and quality of life for a post-encephalitis population may be lower than other similar populations such as those who experience traumatic brain injury and stroke.

In 1999 a study by Moorthi et al. discovered that although rates of recovery vary for encephalitis survivors, it is generally less than for those affected by stroke or traumatic brain injury. More recently in a study using a measure called the European Brain Injury Questionnaire (EBIQ – Teasdale et al., 1997), Easton (2012) established that the depth of problems across nine domains (Somatic, Cognitive, Motivation, Impulsivity, Depression, Isolation, Physical, Communication and Core) for people affected by encephalitis are greater in nearly every case than for some other forms of acquired brain injury (traumatic brain injury, cardiovascular accident (stroke) and other miscellaneous causes). Severe cognitive deficits were noted by Pewter et al. (2007) along with abnormally high levels of distress among participants with significant levels of depression, obsessive compulsive behaviours and anxiety.

Similarly when quality of life has been considered in this population, it has been found

that it can be not only lower than the general population but also lower than for other comparable neurological conditions, for example stroke and traumatic brain injury (Stapley et al., 2008). More recently a similar study conducted by Ramanuj et al. (2014: p. 6) provided “compelling evidence that in addition to the significant mortality and morbidity associated with encephalitis, the illness has long-term adverse effects on quality of life for the majority of survivors”.

This growing literature is beginning to provide evidence that might suggest outcomes and quality of life for people in a post-encephalitic population can have a depth and gravity not experienced in the same way as other brain injury population groups. In addition encephalitis is a complex condition that most people have not heard of prior to it affecting their life. This means there is little collective understanding about the condition in the general public and indeed in professional circles, in the same way that there is for other conditions, for example motor neurone disease, meningitis, cancer, and so on. In turn people may experience a lack of understanding of, and empathy for, their difficulties.

Patients recovering from other brain injury-related conditions such as traumatic brain injury and stroke are often in hospital longer due to orthopaedic and other physical deficits. Resultantly their more ‘hidden’ disabilities such as memory, executive functioning and pain for example, are identified during this time and referrals to other services such as neuropsychology and pain clinics are made. Not so for the encephalitis patient, who rarely experiences physical problems and who is often discharged as soon as the acute phase is over. In some cases referral criteria for statutory services may be biased, catering for more commonly occurring conditions such as traumatic brain injury and stroke.

What we have, therefore, is a condition which is complex, about which there is little understanding and patients who are often discharged quickly and without follow-up. In many cases it is an acute illness, which if the patient survives, is often superseded by acquired brain injury. In particular the acute illness is not in itself the start of what is expected to be a chronic condition. It is only as the injury to the brain emerges in some people over time, and as their awareness and acceptance of it begins, that having had encephalitis can be said to have resulted in a chronic condition. Where people die it is likely to be quick and unexpected for the families. In more rare forms of encephalitis people may experience a long and lingering death, which may have some similarities to other chronic (but unrelated) terminal conditions (for example CJD – Creutzfeldt-Jakob Disease).

Whether these variables contribute to experiencing a poorer quality of life remains unknown. Taken altogether and with the experience of a sudden-onset, life-threatening condition, it could explain why people turn to stories in order to understand and make

sense of their condition. Patient narratives have been a feature of medicine throughout history. It is useful to briefly consider here the historical context in which encephalitis narratives exist.

Encephalitis and people's stories

Frank (1997: p. 21) suggests: "Published stories also have a particular influence: they affect how others tell their stories, creating a social rhetoric of illness". In other words the way people tell their stories may be influenced by their reading of stories and this in turn may influence how people perceive themselves, their loved ones and the impact their condition has upon their daily life. The Encephalitis Society staff observe a steady stream of people (those affected or family members and friends) who send in their 'stories' with the intention they be shared with others, perhaps in newsletters or on the website. In many cases the stories follow a familiar pattern: who they were before they became ill, what happened to them (often this section requires the observations of family and friends) and who they are now (post- encephalitis/brain injury). The content of these stories can be moving and heart-rending; confusing and chaotic; bleak and desperate; at other times triumphant, hopeful and inspiring.

Research into chronic and long-standing illness has suggested that narratives play a potentially important role in enabling (some) people to make sense of what has happened to them. Frank (1997: p. xi) states that "The ill person who turns illness into story transforms fate into experience". Frank (1997: p. 13) also raises the issue that "pressures on clinical practice, including the cost of physicians' time and ever greater use of technologies, means less time for the patients to speak. People then speak elsewhere". This combined with other social phenomena such as our increasing geographical dislocation from extended kin, rising use of televisions, computers, gaming devices and other technologies, may all have resulted in reduced opportunities where people once would have spoken and been heard. It must also not be forgotten that the social circles of those affected by encephalitis often decrease dramatically as a result of the limitations they are left with. As already described, relationships may break down, friendships are lost, and employment and colleagues may become a distant memory for some. Personal narratives may therefore play an increasingly important role for those who read and write them. Understanding narratives in their historical context is important in order to appreciate their meanings for different audiences (for example patient versus physician) and how these have changed over time. During the eighteenth century the patient's story was seen as central to the consultation because this was a time before the development of many medical tests and equipment: the patient's story was often all the evidence the diagnosing physician had. Emerging



industrialisation and technologies were, however, in part, responsible for moves away from the importance of the patient's narrative to a biomedical model where the ability to measure physical capacities and strengths were considered the 'gold standard' in terms of diagnosis and treatment: the body became separated from the mind; socio-economic and environmental causes of disease and ill-health were disregarded, and a person's experience of ill-health and disease, along with the meanings they attached to them, were superfluous (Bury, 2001). Disease therefore was considered to be located solely in the body, independent of experience, and patients' accounts of their symptoms were considered to confuse diagnostic processes (Porter, 2003).

A broader interest in patients' narratives has however been re-emerging in recent years because of a shift in acute, life-threatening (often infectious) diseases to more complex chronic states resulting in changes in how they are understood, greater awareness of the wider implications of living with long-term disability and a cultural shift towards self-care and self-management of long-term conditions (Cooper, 2004; Nettleton, 2006). What patients think and say therefore has become more important. This is mirrored in our experiences of encephalitis over the last four decades. The mortality rate of herpes simplex encephalitis (one sub-type of encephalitis) was in the region of 80 per cent; however, with the introduction of Aciclovir (an antiviral agent introduced to the market in 1984), mortality has reduced to 10–30 per cent (Chaudhuri and Kennedy, 2002). Consequently, more people are surviving. Not only are they surviving but they also have to live with the consequences of the damage to their brain. As reported earlier people often report a sense of loss in terms of their skills or capacity, and relationships, as well as describing a "loss of sense of self" following their illness. It is perhaps these losses that contribute to the need to better understand and make sense of their experience or those of a family member. Narratives are one way of achieving this, and are therefore not only a tool which can be utilised by those affected by ill-health, but which can also be useful in supporting healthcare professionals to better understand how people experience their condition.

Conclusion

Encephalitis is, as I have shown, a complex syndrome with variable outcomes and severity. It is a disease considered acute but which, in fact, has long-term consequences and implications for those who sustain injury to the brain as a result. This is further complicated by the hidden nature of brain injury and the lack of a collective understanding for a condition most people have never heard of. These factors often result in a state of confusion for those involved and those supporting them, as well as changes to how people see themselves and feelings around who they once were and

who they are now. Narratives have had a long history and may offer those affected by encephalitis one way of making sense of, and coping with, their condition.

Key messages

- Encephalitis is often an acute illness, and a medical emergency.
- Encephalitis may be caused by infection or by the person's own immune system.
- Brain scanning and lumbar puncture are key investigative procedures in diagnosing encephalitis and determining its cause.
- Acquired Brain Injury (ABI) can be a consequence of encephalitis. Epilepsy can be a consequence of encephalitis, along with fatigue, memory problems and differences in personality, behaviour and emotion.
- There are in the region of 6000 cases of encephalitis in the UK, and an estimated 4.25 million worldwide, each year.
- Some forms of encephalitis are vaccine-preventable.
- Brain injury is a hidden disability.
- Outcomes and quality of life post-encephalitis may have a depth and gravity not experienced by some other brain injury populations.
- Narratives are one way people might better understand, and make sense of, what has happened to them post-illness.

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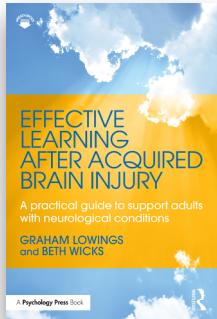
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Understanding Neuropsychological Assessments

Chapter 5: Understanding Neuropsychological Assessments



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The purpose of this brief chapter is to provide the reader with an overview of what to expect in a typical neuropsychological assessment. In the next chapter we will then show how this information provided by the neuropsychologist can be used to inform the Individual Education Plan (IEP). We include descriptions and explanations of the areas likely to be covered in a neuropsychological assessment and, with the use of examples, show how this can be used to produce a cognitive profile of the person. This subsequently will be used to inform the IEP and the strategies to assist the person with ABI to learn.

The purpose of the neuropsychology assessment can be varied. It may have been written to inform rehabilitation or be a follow up to a previous assessment in order to determine if there have been any specific changes to the person's presentation. The following areas are likely to be covered in the report.

- Reason for the assessment.
- Description of the ABI or other neuropsychological condition.
- Background information/history:
 - education and occupation
 - medical/psychiatric information
 - previous neuropsychological or cognitive assessments
 - current situation
 - other observations.
- Specific areas assessed.
- Effort.
- Result of the assessment.
- Implications arising from the results.
- Recommendations.

A neuropsychological assessment can take a number of hours to complete and may have been undertaken over a period of days. This will have provided a comprehensive assessment of the person's cognitive strengths and weaknesses and, specifically, the impact the ABI has had on their functioning.

We will now take each of these areas in turn and provide descriptions of how they are likely to be presented in the report.

Reason for the assessment

The person will have been referred for a neuropsychological assessment for a specific reason. Typically this will have been to aid diagnosis and to inform future treatment and rehabilitation. The assessment may also have been requested as part of a legal process, for instance to provide evidence of the level of disability following the ABI. At some point in the future, the authors would like to see neuropsychological assessments conducted specifically for the purpose of informing a person's IEP. Some aspects of the neuropsychologist's report may not be directly relevant to the educator or facilitator but we include explanations here so the reader can see how their role fits in with the wider care of the person.

Description of the ABI or other neuropsychological condition

In Chapter 1 we provided a description of the brain. In this section, the neuropsychologist will have described which parts of the brain have been damaged as a result of the ABI. In doing so, there may be references made to medical opinion, the results of imaging such as CT and MRI scans and a description of how the person presents in general terms and how their injuries have impacted on activities of daily living, often shortened to ADLs.

Background information/history

Typical subjects covered here will include information about the person's education, employment and home circumstances. If relevant, details concerning friends, family and carers may be included here. There may also be a description of how the ABI occurred and any information regarding previous injuries or diseases that may continue to have impact on the physical, psychological or cognitive performance of the individual.

Areas assessed

Pre-morbid ability

'Pre-morbid' is a medical term that means prior to the illness, disease or injury and this is important detail. Knowing how a person performed before their ABI allows for sensible rehabilitation targets to be set and can be very relevant when planning educational programmes. If we underestimate the person's previous abilities, we will not set demanding enough targets for the individual and we may miss opportunities to maximise their potential. If we overestimate the person's abilities, we run the risk of



setting targets that are far too demanding and which are likely to cause unnecessary stress and demoralise or demotivate the person. The optimum aim of rehabilitation is to return the person back as near as possible to where they were physically, cognitively and emotionally before they had their ABI. Pre-morbid ability is an estimate and not a factual measurement. It is based on a combination of factors, including educational attainment, employment history, observed behaviour and the completion of a psychometric test, most commonly a form of single word reading test. For our purposes, this section will provide you with detail of the person's previous educational attainment and give some indication of their strengths, preferences or potential before their ABI.

Cognitive areas assessed

Though not an exhaustive list, the following areas are most likely to appear in a typical neuropsychological report:

- general intellectual ability (IQ)
- attention
- memory
- executive function
- perception
- language
- processing speed.

In Chapters 4 to 9 we describe these and other cognitive areas likely to be impacted upon by ABI. We include descriptions of the typical difficulties that can be experienced within each cognitive area and how, with the aid of appropriate strategies, these difficulties can be overcome allowing effective learning to be made.

This is a brief description of the type of tests neuropsychologists use to assess ability in those various cognitive areas. Neuropsychologists have many tests to choose from and generally will have their preferred test for each of the cognitive domains.

General Intellectual Ability, if reported, will most likely be the result of the administering of the Wechsler Adult Intelligence Scale, version 4 (WAIS-IV). This involves the completion of ten individual subtests, which result in a measure of intelligence across a number of subject headings, commonly referred to as 'indices', namely:



- Full Scale IQ
- Verbal Comprehension
- Perceptual Reasoning1
- Working Memory
- Processing Speed

Tests for attention are based around one or more of the senses (often referred to in reports as ‘modalities’), most commonly the ability to concentrate or attend to auditory stimuli, or visual stimuli. The test may require the person to listen to something, perhaps count a number of beeps heard in a given time or to find and mark as many occurrences of a particular symbol within a complex picture in a given time (a measure of sustained attention). Occasionally, the tests involve a combination of both modalities to see if we can ignore one stimulus whilst concentrating on the other (a measure of selective attention) or to switch from one modality to another (a measure of attentional switching, demonstrating cognitive flexibility).

We describe in Chapter 6 the complexity of what we term as memory. A comprehensive memory assessment needs to consider all these complexities. If the memory deficit is clearly located in one area, the neuropsychologist may decide to measure memory performance in that discrete area alone. Conversely, they may decide to complete a more comprehensive assessment of memory, which would be most useful for our purposes as it will involve the identification of the person’s aspects of memory that are working well as well as those aspects that are causing them difficulty. As you will see from Chapter 6, we can use these good aspects of memory as part of a set of strategies to compensate for those memory weaknesses. Typical tests of memory include being given lists of words or a short story to remember and then being asked to recall the information after a given time. For visual memory, the person may be asked to replicate a line drawing from memory. Similarly, they may be presented with a set of photographs or drawings and asked later to select which items they had seen previously, from a larger album. They may be asked to observe some behaviour and later in the assessment, to repeat that behaviour themselves.

In Chapter 7, Executive Function is described. You will recall that this term is used to consider a person’s ability across a range of tasks including: planning, problem solving, initiating action and self-monitoring or evaluation of actions. Typical tests of executive functioning include: finding your way round a route or map, solving a practical problem or completing a number of tasks within a given time frame, such as writing a letter, doing some arithmetic and making a list of items seen.

As there are so many activities that we cluster under the term ‘executive functioning’, it is important for the neuropsychologist to be clear about what specific aspects of executive functioning they have examined and the impact any executive deficits are having on the person’s ability to function in daily life.

We briefly discussed verbal and visual perception in Chapter 4. Significant verbal perception difficulties will be apparent during conversations, including during the administration of neuropsychological tests. Typical tests for visual perception include ‘cancellation’ tests, where the person is required to cross out a target shape from an array of similar shapes. Persons with hemianopia will cross out targets on only one half of the paper, not seeing the other half. Other visual attention tests include being required to create set patterns with wooden building blocks and to complete a series of visually presented puzzles.

In Chapter 9 we describe some of the various differences with language that can be experienced by those with an ABI. The person may have been asked the meaning of specific words, how different words may be similar and questions of general knowledge. Their word-finding ability may have been tested by asking them to say out loud the names of items presented visually, often in the form of a line drawing or a photograph.

The cause and nature of a person’s vocabulary difficulties may be complex. It may be they have forgotten or are unable to retrieve the names of items or these words have been lost to their vocabulary since the ABI or it may be a sign of others factors. A test of verbal fluency may have been used to assist in identifying the level of difficulty and possible cause(s). Verbal fluency is tested by asking the person to say out loud words beginning with a specific letter or within a specific category, such as boys’ names. The person may be asked to name items within two categories, alternating between them. Where the main difficulties occur with words starting with a specific letter or words within a specific category or a combination of both, the results will have provided the neuropsychologist with strong indicators of the cause of the difficulty, whether it be trauma or disease. As the verbal fluency test is also a measure of cognitive flexibility you may also see this reported under Executive Functioning.

From conversations with the person, difficulties affecting speech, such as dysarthria and aphasia, may be readily apparent but other language problems may be less obvious initially. If the person has language and communication difficulties there is likely to have been an additional report from a specialist speech and language therapist. Like the neuropsychologist, they have their own specific psychometric tests to support their assessments and they will have explained in their report how the person’s language difficulties are impacting on their functioning in daily life and hopefully will have included strategies and exercises to help the person overcome and improve on their

language ability.

As discussed in Chapter 8, the speed at which people process information can have a significant impact. This may have been measured by asking the person to read simple sentences, to write simple sentences quickly or to perform simple arithmetic calculations within a specific time. More commonly the person will have been asked to complete one or more of the following tasks within a given time frame:

- Cross out a target shape from an array of similar ones.
- Decide if one of two shapes is present in a line of similar shapes.
- Using a key, where numbers 1 to 9 each represent a unique shape, write or say out loud the corresponding number of the shapes randomly presented. A task requiring a verbal response is useful for those whose ABI has caused physical problems, making hand-writing difficult. This may be presented in the reverse, with the number provided and the person having to respond by drawing the corresponding unique shape. This is useful where the person is experiencing speech difficulties.

The more of each task the person successfully completes, within the given time, the faster their speed of information processing.

Mood

In Chapter 4, we included a reference to mood. The neuropsychologist is likely to have considered the person's mood in terms of motivation, anxiety and depression. Their opinion may be based on observation and the views of others, such as partners. Most likely, the person will have been asked to complete one or more questionnaires, which give indications as to the person's level of anxiety and depression. Less common but still possible are questionnaires completed relating to motivation and irritability and the ability to control anger. All of which is useful information when considering how to manage behaviour.

Effort

This can relate to a number of issues. It may relate to motivation and the level of commitment and enthusiasm the person has demonstrated during the assessment. There may be reference to and information regarding executive functioning if issues such as poor initiation and monitoring have contributed to what may be perceived as lacking effort.

The majority of patients seen for neuropsychological assessment give of their best and,



therefore, the results reported are a true reflection of the subject's abilities, deficits and difficulties. However, a small proportion may not give their full effort for a number of reasons. It is for the neuropsychologist to consider this and identify to the reader should level of effort be in doubt.

Result of the assessment

Raw scores, scaled scores and other statistical information may also be included, though these tend to be provided only when the reader is likely to be another psychologist to ensure the data is not misrepresented or misunderstood. Commonly, qualitative statements will be made (see Table 12.1) and if statistics are quoted they are most likely to be percentiles. Basically, a percentile shows how the person performed when compared to their peers. For example, if it were reported that the person performed at the 37th percentile in an attention test this indicates that the person performed as well or better than 37/100 people.

Table 12.1 provides details of the qualitative statements commonly used in neuropsychological reports, together with the percentile ranges for the normal population to which the statement refers.

Table 12.1 Percentile range and matching qualitative statement

<i>Percentile range</i>	<i>Qualitative statement</i>
98th and above	Very superior
91st to 97th	Superior
75th to 90th	High average
25th to 74th	Average
9th to 24th	Low average
2nd to 8th	Borderline
Below 2nd	Extremely low

Implications arising from the results

In this section, the neuropsychologist should provide his or her professional opinion as to how the results will be reflected in the patient's ability to function in everyday life, at home and at work, if applicable. There may be comments to compare how they performed before the ABI and how their performance has changed since. It is important to stress that the neuropsychologist should highlight the strengths identified by the



assessment as well as the difficulties the person now experiences. Future strategies used to help the person will focus on making effective use of these strengths as part of an overall package of care aimed at managing or compensating for their difficulties.

Recommendations

The neuropsychologist's recommendations may include:

- Referrals to other health care professionals to address specific issues identified in the assessment – for instance, referrals to Ophthalmology or Optometry for visual difficulties identified.
- Advice to the person or to his or her carers regarding support services they may wish to access, such as the charities Headway and the Stroke Association, or Different Strokes for the younger stroke survivor.
- Guidance may be included on how to use specific strategies that the individual or carers can employ to help with improving functional ability in activities of daily living. These may contain elements of the strategies we have described in this book.

In the next chapter we describe how we can use the information provided in the neuropsychologist's report to establish a cognitive profile and ultimately an IEP to enable the most effective learning experience for the person with ABI.

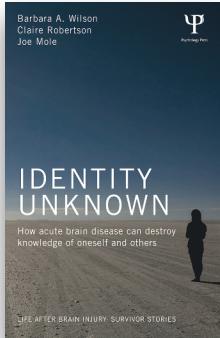
Note

1. Perceptual Reasoning is a category of reasoning skills that includes non-verbal fluid reasoning, spatial processing and visual perception. Simply put, making sense of what we see.



Strategies that have helped

Chapter 6: Strategies that have helped



The following is excerpted from *Identity Unknown* by Barbara A. Wilson, Claire Robertson and Joe Mole. © 2015 Taylor & Francis Group. All rights reserved.

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Barbara has asked me to talk about my life now. I have come a long way since I kissed the cauliflower thinking it was Ed! Before I start with all the practical chaos, I just want to say that I am feeling much more secure about my life now as I am using many strategies to guard and record my memories.

I have rehabilitated well (not recovered). I am left with prosopagnosia, short- and long-term memory loss, risk of further fitting, reduced speed of thought and poor organisational abilities. I get tired easily and have regular headaches, am anxious most of the time with low self confidence. I am sensitive to sudden loud sounds, sudden unexpected movements and bright lights. I need to avoid situations with flashing lights or high pitch music to prevent me having another fit. I am lucky to have warning signals and the physical ability to alter the circumstances around me. I have anosmia (loss of sense of smell) but am very happy I can see the daffodils even if I can't smell them. And I have children at home who tell me when I have left the gas on! Taste wise, things are improving and I am managing to try and enjoy a few sweet things without them tasting metallic. A bit of lemon, mint or ginger can really help the chocolate be almost nice. I feel I must be mending some connections over time. I am usually quite stubborn, trying very hard to manage all my own expectations and hopes for myself without bothering others so as to hang on to some fragments of my own self-esteem. But I know that I am unsafe at times and have learned the hard way—as we all do. Life is not a dress rehearsal and I am trying to relearn about Claire Robertson from all the other people who know and understand much more about her than I do.

Let's visit a typical day for me. The alarm goes off. I awake on my side of the bed with the man I believe to be Ed. Then there's the school run—chaos for everyone! I have shared this palaver with all four of our kids and their friends over the years, but now that I can't recognise them or any of their friends or remember who needs to go where—it is all a bit of a scare. I feel very shy to ask my friends' children who they are, again, and where to take them, again, and it can be very embarrassing for my children to have Mum having to ask everyone again, including them! Even when they have told me, I can't remember a minute later and need to ask again. Over time I have learned to keep calm and try to be friendly, however confused and fed-up they get to be, but I very much want them to realise that I do care very much about them all individually, but through the years of "teenage rampage" it's been a very big learning curve for all of us.

OK, that's the school run done, I can go and do my exercise class. I have time now that I am not able to work. I've been to aqua aerobics, my serious prosopagnosia challenge, I get out into the car park and encounter other people parking and going in. They may or may not know me, speak to me, or wish to ignore me, I drive home, and whether I risk the same

difficulties at Sainsbury's (the supermarket) is another question. Do I risk it? But I need to do the shopping, my family rely on me to manage this. Before I know it, I am being hugged in the fridge department by somebody very pleased to bump into me, and they seem to know me! Who can they be? I go to the till with the lady working there, who happily greets me like an old friend as well. I have learned to respond to this in a friendly way and keep packing the bags and reading labels rather than making eye contact with anyone else. I say "Bye" very certainly and escape to the car without looking at anybody, in case I get another "Hello Claire". I think I must look very worried to people, who seem to warm to me as if I'm a frightened child.

Home, where the post is on the doorstep and some of it is addressed to me. Who is it from? I try to assess the writing and the postmark and work out if it might be something I am expecting. I may open it up and find a name "Joe Bloggs". I don't recognise the postmark and the written piece is small and all scrunched up and becomes impossible for anyone else to read. But it says "Hi, Claire", as if someone is there, and the more I decipher, the harder it is to understand who has kindly sent me this card. My first visit is to my A-Z Identity Book, which will help me to realise who this is from. I look up their name, helpful if I can work out the surname but each page becomes a discovery in itself. It tells me which Linda it is from and how we know each other. I have a rush of sadness with lost memories but a warm secure feeling that our friendship is still building, Building! That says it all—the "wall of friendship" becomes stronger with every meaningful, understood contact. And the great thing about the written word is that I can keep it. It has a security of its own Strategies that have helped and I store it in the right section of my Friendship File. It feels as if our friendship is still intact and I can revisit it whenever I like, and, hopefully share it with Linda too.

Phone calls are nowhere near as easy as the written word. I still dread answering the phone and rarely use it for socialising if I can avoid it. The spoken word goes too quickly for me. I can't remember it in time to allow me to appreciate what is being said. E-mails and phone texts are not so easy to store and feel secure about. Although I know that printing off a computer is expensive and not the best thing for the environment, I find myself using the printer and storing words in a desperate attempt to keep control of my own understanding of what has happened in my life and relevant aspects of other people's lives. Texts get scribbled and stored, in another "waste" of paper, but they feel like a more secure way of accessing parts of my memory. How to get somewhere to share life seemed to be so easy and taken for granted before I lost my memories and confidence in my ability to navigate myself. Now that I have been able to re-use the written directions over and over again, I use this less and less now and find myself following my nose and sense of direction. My visual memories of these written words and maps have made all the difference. I have made myself a "Present File", recording who has kindly given me which gifts, as I felt heartbroken knowing that I have some lovely items but no idea which kind person chose them for me.

Choosing presents and using them is all part of the fabric of friendship and love—it also shows that we choose to care about that person's needs and feelings by giving them lovely things. I have felt much more secure about my gratitude now that I am making written records. I can even use the right mug when my mother-in-law comes round for tea! And wear that lovely top she always reminds me that we chose together. Caring needs protecting and, in order to feel caring towards others, I need to store many memories to ensure that I react to them correctly. I have had many different files and lists. One day I came across an A4 arch file with photos of alphabetti spaghetti all over it, with the letters spelling "Brain Food". It was for me! In it, I keep my own important information files and documents, which I need to keep safe in order to continue to cope with being me.

Writing to people isn't easy, as I need to evoke my own thoughts and feelings in order to make meaning out of what I am writing about. It's like coming into a random chapter in a book you've never read. You open the page, expect to know what's happened, what's happening and guess where it's all going! I gather together all my friendship files and pieces of writing I've stored and these help me to feel that I am more able to write to this person. Thank goodness for pen and paper, and how fortunate I am to be able to see.

Over ten years of troubles I have come a long way with managing my difficulties as best I can without being able to recognise people by their faces. I am going to try to explain how things have improved.

The following are the strategies and devices I use now. Current memory system:

- *Filofax—excellent resource but in a big muddle*
- *Daily To Do Diary—now using the following method:*
 - *As and when*
 - *If ever*
 - *To do*
- *White board on fridge*
- *Family diary*
- *Notebook in camera case to make notes*
- *A-Z identity jig—the best thing ever!*
- *Friendship Book and Friendship File, a great new device but well behind where I want it to be*
- *SenseCam work and diary writing—very helpful*
- *Jobs to do list, shopping lists and reminders*



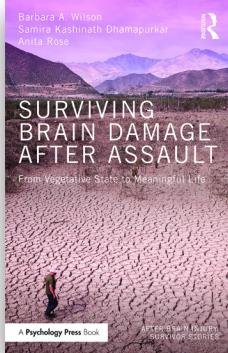
- *More organised boxes with named files*
- *New photo organisation box—in chaos*
- *Book of written directions*
- *Files for paperwork that needs doing, labelled to remind me what to do with them*
- *Recipes that have worked notebook*
- *Meal planning and shopping lists*
- *Notepaper by phone.*

And now I have scribbled all this lot, I need to re-read all my rehabilitation advice and use it again to get even better organised.



Why did Gary do so well?

Chapter 7: Why did Gary do so well?



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When Gary was first seen by the current authors, none of us predicted he would do well. He was in a vegetative state and remained so for a long time. Only a minority of people regain full consciousness after such a long period, and very few recover as well as Gary. It is well recognised that patients who are in a VS following anoxic damage do less well than those whose VS follows TBI (Giacino & Whyte, 2005; Guérin 2005; Kotchoubey 2005). Anoxia appears to result in irreversible neuronal loss. Andrews (2005) lists five conditions which should be provided to promote optimal recovery for patients with profound brain damage. These are:

1. Provide the optimal environment.
2. Prevent and treat secondary complications.
3. Include in treatment physiotherapy and the other “therapies”, medical, psychological and technological.
4. Support the family.
5. Modify the environment, including regulating the amount of stimulation.

It is certainly the case that all these conditions were met in Gary's rehabilitation programme.

We know too that people denied rehabilitation do not do well (Elliott & Walker, 2005). Rusk and colleagues (1966) followed up 25 survivors of brain injury 5 to 15 years after they were considered unsuitable for rehabilitation. Five had died, and of the 18 patients still alive, there were “hundreds of incidents of infections and respiratory complications. Contractures were evident in all patients even those initially without contractures” (Elliott & Walker, 2005, p. 482).

We asked several people why they thought Gary had done so well. Wendie, his mother, considered it was tender, loving care, perseverance and not giving up, together with the staff and people at the Raphael Medical Centre. She recognised that most people do not have the same rehabilitation opportunities as her son. She told us that if he had gone into a home, he would probably be sitting hunched up in his wheelchair all day.

On January 14, 2015, the director of the RMC, Dr Gerhard Florschutz, was asked why he felt Gary had done so well. This is a transcript of the interview.

BW: Why do YOU think Gary did better than many people predicted?

GF: Well, it's very difficult to say, but if you look at the pathway, the history of his recovery, there was a pronounced difference after his cranioplasty, and I think since the cranioplasty he made progress that he wasn't able to make before. Then, of course, there was the input of the interdisciplinary team working very intensively with him, not just the physiotherapists but all the other therapists working with him, occupational

therapy, neuropsychology and so on. That made a difference. The synergy of these various therapies really makes a difference.

BW: Have you seen other people like Gary? How unusual do you think he is?

GF: Well, he's very unusual. When one looks at his history, at the degree of his injury and the trauma and the length of his coma, I would say it's quite unusual. I mean we have had one or two other cases of a similar nature that have made an almost total recovery. Again, it's unexplainable why that happens.

BW: Do you think his family had anything to do with this?

GF: The mother most certainly was very important, and one can't underestimate her input. She was so motivating to him, pushing him and pushing herself as well, in a most positive way, that was very helpful.

BW: The other thing about Gary is that he failed to show improvement for such a long time that in normal circumstances, he would have been sent to a nursing home, and I feel that if that had happened he would have ended up very contracted (having permanent shortening of muscles or joints). Is that what you feel?

GF: Yes, that is really a lesson we have to bring to the attention of the funders. In many cases, they have to give space and time for recovery to take place. Then, in economic terms, the benefits would have been obvious. He would have been a cost to the health service to the end of his life. Now he will cease to have health funding needs very shortly. So from that point of view, neurorehabilitation is cost effective. He is a good example. That is really the problem that this medical orthodoxy has: that declares that after 6 months you are in a "persistent vegetative state" and after a year you are in a "permanent vegetative state" is nonsense. There are so many examples where people make recovery and improve long after that time. What we have to develop is a system where we can detect possibilities for recovery. How can we detect early on who can make a better recovery? That is a real research question. That is something we really have to work on.

BW: Well, that's an interesting one, but I don't know that it's possible. You know that Agnes Shiel, in her PhD in 1999, found five different patterns of recovery. One group remained vegetative while another group remained flat for a long time and then recovered. Gary would fit into this group. Then there were other patterns. The point of that study was that you couldn't predict which ones were going to do well in the first few months.

GF: With Gary, in one way, you could have predicted he would do well because of the cranioplasty. There are enough data now that, after a cranioplasty, enough people make a recovery, some more than others. This is one thing we can look at. [See Chapter 8 on

cranioplasty in this book.]

BW: The other thing about cranioplasty is the timing. Some people, for example in Germany, think you have to do it quickly, whereas in the UK people tend to wait. Is it just money or risk of infection or what?

GF: The risk of infection is there anyway whenever you do it. If I'm being cynical, I think it's because they are waiting for them to die so they don't have to pay for it. It's almost as cruel as that. It makes sense to do it quickly because if you really want to encourage plasticity of the brain, you have to have a properly enveloped skull to function better. That is often the reason why people recover better after a cranioplasty.

BW: How close was Gary's waking up to the cranioplasty? It was quite a while afterwards, wasn't it?

GF: Quite soon I think.

[The cranioplasty was carried out in August 2012, and Gary emerged from the MCS in April 2013, 9 months later, but he did appear to be more awake after the cranioplasty.]

Anita Rose: I think there were some changes in his behaviour, he became more alert, and that was quite soon afterwards. [In fact, Gary emerged from the VS into the MCS soon after the cranioplasty.]

Samira also reminded us that Gary had remained physically well while at the RMC; he had not developed infections; which can often pull people down. There had been no further falls; his medication had been regularly reviewed; he had relatively few seizures, he never developed pressure sores and good nutrition and hydration had been maintained. Thus, his physical state had been the best it could have been, thus encouraging any recovery to take place. Wilson, Graham and Watson (2005) are among others who stress the importance of good physical care and the need to deal with treatable problems during the VS. They mention how patients who are heavily sedated can show improvement once the medications are stopped or adjusted.

Our colleague Lindsay McLellan, a neurologist and former professor of rehabilitation, when asked why he felt Gary had done so well, sent the following comments by email on February 10, 2015:

1. I think the mechanism of the initial injury was significant in that his head was likely to have been relatively stationary when hit (thus unlike a motorist travelling at speed); moreover the blow fractured the skull which thus absorbed some of the force of the blow and caused less catastrophic injury to the brain than initially thought.

2. Hydrocephalus is a very significant complicating factor; I suspect that current methods of assessment of the need to reduce intracranial pressure in an individual are



insufficiently precise. The standard way of evaluating clinically whether a permanent shunt is needed to reduce the pressure is in my opinion lacking in subtlety and is far, far too brief, so that some people who would benefit from a permanent shunt are not given one. Once excess pressure is relieved, it may be several weeks or even months before clinical improvement appears and further improvement may then continue for 2 to 3 years and confound earlier predictions. (I have seen one such striking case similar to Gary's, that occurred in response to long term input from his family following decompression). Sometimes it takes a surgical procedure such as replacement of a bone flap to trigger the process of long term decompression and I suspect hydrocephalus was a significant factor in Gary's case, given his high level of eventual cognitive recovery. [In support of McLellan's view, Pickard and colleagues (2005) suggested that in vegetative patients, hydrocephalus is less likely to present as raised intracranial pressure and more likely to present as failure to improve.]

3. In addition, I am sure rehabilitation is crucially important. I think that a small proportion of individuals who are clinically unresponsive are actually aware as if in a dream but lack awareness of their ability to respond or comment, as though stuck in 'watching TV' mode. They can eventually be encouraged out of this by a combination of specific physical arousal (as occurs during physiotherapy) and personal and cheerful appeals to respond which may need to be theatrically exaggerated (rather as parents intuitively adopt when encouraging young children). However, such responses may initially be very short lived, fatigue rapidly and be easy to miss. So frequent short inputs from someone sensitive to subtle changes are more effective than relatively infrequent 45 minute sessions. By contrast, I suspect that simply leaving people continuously attached to headphones or parked next to interminable TV induces a state of withdrawal from stimulation and thus delays recovery.

Another colleague, Agnes Shiel, the senior author of the WHIM, also sent an email on February 12, 2015, in which she wrote:

"Leaving aside the obvious explanations such as medication, nutritional status etc., which will have been considered, there is no obvious answer. It may be due to a combination of factors including careful assessment which identifies subtle signs of improvement leading to appropriate types and intensities of intervention. Another possible factor may be persistence in intervention – in many cases, rehabilitation is time limited and, where there is no obvious response after a given time, the focus changes to management and long term care. Perhaps some people with potential for improvement require persistent intervention over time in order to begin responding and simply do not get this opportunity, as Gary did here at the Raphael. A second factor may be readiness to respond and it could be that, serendipitously for some people, this readiness coincides with their access to rehabilitation. However, for many people with a

Disorder of Consciousness, once the initial assessment and rehabilitation is completed, they are moved to continuing care where regular intervention if available at all is of a much lower intensity. Because no change is expected, the person is not challenged and therefore no change occurs. This case illustrates that for some individuals (albeit probably a minority) significant recovery is possible and can be maximised in a favourable environment".

Borthwick (2005) reminds us that patients described as being in the permanent or persistent VS **do** sometimes recover and that a paper on treatment decisions for VS patients from the *British Medical Journal* (Andrews, Murphy, Munday, & Littlewood, 1996) said the reported recovery of some of these patients was a cause for concern. Kotchoubey (2005) explains that VS patients may have islands of cortical activity but that these are disconnected from one another. In other words, there may be "lack of activity of large distributed cortical networks despite intact cortical activity" (*ibid.*, p. 349). Wilson, Graham and Watson (2005) highlight the fact that there are some VS patients who show isolated behavioural fragments. Katchoubey (2005) tells us that there are two subtypes of VS, one with relatively intact thalamo-cortical links and one without. Gary presumably belonged to the former subtype. Rappoport (2005) also describes two subtypes of VS, namely "vegetative state" and "extreme vegetative state". Once again, Gary would appear to be in the former subtype.

Let us return to the patterns of recovery mentioned (Shiel, 1999). Using the WHIM, Shiel identified five different patterns of recovery in survivors of severe brain injury: (1) little evidence of change; (2) slow, steady progress; (3) rapid progress with a quick plateau; (4) initial rapid recovery, then slow progress; and (5) little if any progress at first, then steady progress for a long period. These patients, however, were in the acute stage. What about patients who remain unaware for many months? Dhamapurkar, Wilson, Rose and Florschutz (2015) wanted to know if the same patterns of recovery were seen in chronic patients using the same measures as Shiel (1999). They looked at 29 patients surviving severe brain injury who were in a VS or a MCS for several months. All were assessed regularly with the WHIM (Gary was one of the patients included in this study). Three of the participants had died. Of the remaining patients, 17 showed little evidence of change (Shiel's group 1). Three showed gradual improvement over time (Shiel's group 2). Two patients improved and then plateaued early (Shiel's group 3). Only one patient rapidly improved (Shiel's group 4). The final three patients were slow to start and then showed a marked improvement (similar to Shiel's group 5). Gary was one of these. The authors concluded that the taxonomy suggested by Shiel (1999) also appears to apply to patients in the chronic phase but with a delayed start of several months. They recognised that the findings need to be replicated with a larger group of patients. Guérat (2005) also mentions different patterns of recovery seen after TBI as measured by EEG. One final point to make here is to remember Rosenblum (2015), men-



tioned in the recovery section in Chapter 3, who says that recovery after long periods of coma is possible if the critical connections between the brainstem and the ascending arousal system are intact. Gary did not have significant damage to the brainstem, and this may have contributed to his good outcome.

To quote Province (2005), “The customarily grim view of severe brain injury resulting in cognitive impairment provides a disincentive for the development and implementation of rehabilitation strategies that could benefit these patients” (p. 269). Gary is an example of how the principle of *never giving up* has led to a good quality of life for one such patient.

