

BRITISH JOURNAL OF
NEUROSCIENCE
NURSING

THE ONLY UK TITLE FOR NEURO NURSES

Encephalitis, a service orphan:
The need for more research and
access to neuropsychology

Ava Easton, Karl Atkin, Elaine Dowell

Encephalitis, a service orphan: The need for more research and access to neuropsychology

Ava Easton is Development Manager, The Encephalitis Society, Karl Atkin is Senior Lecturer in Ethnicity and Health, University of York, and Elaine Dowell is Resource Centre Manager, The Encephalitis Society, 7b Saville Street, Malton, York YO17 7LL. **Correspondence to:** Ava Easton

Encephalitis is inflammation of the brain tissue. It can occur at any age, in any part of the world. It is caused by either an infection (usually a viral infection) or by an autoimmune disease. The initial stage of the illness commonly manifests as serious and acute, often followed by some level of injury to the brain. The degree and severity of permanent brain injury varies among those affected.

Statistics from the USA provide an indication of the prevalence of encephalitis. Beghi et al (1984) reported the annual incidence as 7.4 people per 100 000 and, more recently, Khetsuriani et al (2002) reported 7.3 hospitalizations per 100 000 population. There are no statistics for the UK. However, based on the US statistics, it can be estimated that about 4000 people are newly affected each year. Mortality resulting from infection by the herpes virus (the commonest identified cause of viral encephalitis) is 20–30% with treatment. If encephalitis is left untreated, the mortality rate is about 70% (Schott, 2006).

The aim of this article is to raise awareness of encephalitis and its consequences, inform professionals about epidemiology, and highlight the usefulness of approaches such as neuropsychology in the treatment of those people affected.

ABSTRACT

Encephalitis and its consequences are often misunderstood, not only by the general public, but also by those providing health, educational and social services to those who have been affected. The aim of this paper is to provide a brief overview of the epidemiology and clinical consequences of encephalitis, and highlight the potential of approaches such as neuropsychology in its treatment, with the intention of helping health professionals improve the quality of care received by patients. It is argued that rehabilitation of people affected by encephalitis may benefit from interdisciplinary and biopsychosocial approaches, and that neuropsychological assessment can be a useful tool in recovery.

Key words

■ Encephalitis ■ Acquired brain injury ■ Rehabilitation ■ Neurological disease
■ Neurodisability

Accepted for publication following double-blind peer review 7 November 2006.

Symptoms

Encephalitis frequently begins with a flu-like illness and headache. More serious symptoms follow, hours to days later. The most serious sequela is an altered level of consciousness. This can range from mild confusion or drowsiness to loss of consciousness and coma. Other symptoms may include a high temperature, seizures, aversion to bright lights, inability to speak or control movement, sensory changes, neck stiffness, and uncharacteristic behaviour (Encephalitis Society, 2003).

The range of possible symptoms and their rate of development vary widely. These symptoms may also indicate other illnesses, e.g. tuberculosis, meningococcal meningitis, tumour, subdural haematoma, vascular disease, Reye's syndrome. As a consequence, a specific diagnosis can be difficult (Whitley and Gnann, 2002).

Investigations

Investigations undertaken in hospital are likely to include a brain scan (computed tomography or magnetic resonance imaging) to show the extent of the inflammation; an electroencephalogram (EEG) to determine unusual patterns of brain activity; and a lumbar puncture to identify whether an infection is present in the cerebrospinal fluid. Diagnosis of encephalitis is made where there is evidence of inflammation. Polymerase chain reaction (PCR) testing may prove useful in identifying the more specific nature of the infection in some cases. However, in more than half of cases of viral encephalitis, the identity of the infecting agent is not conclusively determined (Booss and Esiri, 2003; Glaser et al, 2003).

Treatment

Prompt treatment with aciclovir is important. Aciclovir is an anti-viral agent effective against herpes viruses. While not always identified, herpes simplex (the cold sore virus) is a common known cause of encephalitis (Schott, 2006). There is no specific treatment, at present, for other viruses infecting the brain. Other treatments are symptomatic and may include anti-convulsants to control seizures and sedatives to reduce agitation. Intensive care with ventilation may be necessary in severe cases to reduce inflammation. Antibiotics are given to prevent bacterial infections. Post-infectious encephalitis is treated with steroids and on occasion with intravenous globulin therapy.

Acquired brain injury

Nerve cell damage and death is caused by the viral infection, the immune response and by extreme pressure resulting from the inflammation. Some loss of brain function is therefore a probable outcome of encephalitis (McGrath, 1997; Aygun et al, 2001; Raschilas, 2002). In some cases, however, this loss occurs on a relatively small scale, resulting in very minor impairment such as some loss in speed of thinking. In other cases, damage can be extensive leading to significant impairments.

The degree and type of damage varies according to the severity of the infection and the parts of the brain that have been attacked. For example, the herpes virus commonly attacks the temporal lobes, invariably affecting memory function (McGrath et al, 1997). The varicella zoster virus commonly affects the cerebellum, resulting in balance problems (Booss and Esiri, 2003). The damage may be limited to one part of the brain (focal), in several parts of the brain (multifocal) or throughout the brain (diffuse). The greater the number of areas of the brain or the larger the area affected, the more likely are wider-ranging and more complex difficulties. It is the combination of location and severity which determine the pattern of problems that remain after the illness (Booss and Esiri, 2003).

Recovery

The acute phase of the illness can last anything from a few days to 2 or 3 weeks, and occasionally may be longer. The acute phase may be followed by a period of fairly rapid improvement that slows down but continues over the coming months and sometimes over years.

With improvements in drug therapy and intensive care treatments it can be expected that many more people are surviving encephalitis, although statistics are difficult to obtain.

No two cases of encephalitis have an identical outcome. Because present understanding of how the brain works remains limited, the outcome cannot be reliably predicted. Outcome will therefore vary between those who are able to return to their former work and life style (with perhaps only a slight change in their ability) and those left with a profound disability, whether physical, cognitive or both. The experience of the Encephalitis Society suggests that a small percentage of those affected by encephalitis will need to remain in residential care for the rest of their lives.

The consequences of encephalitis

Clinical outcomes

The consequences of encephalitis and their impact on the people it affects (both directly and indirectly) is poorly understood. Lewis et al (2005: 356) state that 'encephalitis is an uncommon and disturbing illness whose cause often remains enigmatic despite extensive diagnostic efforts'. Uncommon it may be, but it is important to understand that those affected (directly and indirectly) are many. It is surprising therefore to learn that incidence rates for

encephalitis are in fact higher than for motor neurone disease, multiple sclerosis and some forms of brain haemorrhage (The Neurological Alliance, 2003).

Raschilas et al (2002) showed that death and disability in people with herpes simplex encephalitis are high. They found a death rate of 15% even with treatment, a severe disability rate of 20%, and that about 51% suffer with mild-to-moderate disability. Raschilas defined disability according to the following 5-grade scale derived from the Glasgow Outcome Scale (*Table 1*).

Cognitive, psychological and physical outcomes

People affected by encephalitis may be left with cognitive, physical, emotional, behavioural and social difficulties (Dowell et al, 2001). Moorthi et al (1999) conducted an 8-year study looking at rehabilitation outcomes following encephalitis and found that not only do rates of recovery vary but that they are less than for other types of brain injury such as stroke and traumatic brain injury. Clark et al (2006) acknowledge that 'it is known that psychiatric disorders are more common in individuals who have an acquired brain injury'. However, an (as yet) unpublished study by Pewter et al from the University of Exeter suggests that, in comparison with other forms of acquired brain injury, general levels of distress are high in people with encephalitis. In particular, Pewter et al have identified a higher incidence of depression and obsessive behaviours.

People may experience marked changes in their view of themselves: they are not quite the person they were before. Severe memory problems may result in a disturbing lack of continuity and order to the person's life, in particular, for those with loss of memory for events before as well as after their illness. Changes in personality, especially those involving loss of control over emotions, thoughts or actions, can be profoundly disturbing. Perceptual or

Table 1. A definition of disability according to Raschilas et al

Grade I	Good recovery, allowing independent life without any neurological impairment
Grade II	Mild disability, defined by the presence of minimal cognitive alterations (speech disturbances, memory, or attention impairment) and/or seizures (partially controlled with anticonvulsant drugs), without consequences for socio-professional life
Grade III	Moderate disability, defined by criteria identical to those of group II but with consequences for socio-professional life
Grade IV	Severe disability, defined by loss of autonomy requiring institutionalization or constant life aid
Grade V	Death
From: Raschilas et al, 2002.	

spatial disorders can present a confusing and bewildering world within which it is difficult to play a full part. For those with reasoning difficulties life may appear at times an insoluble puzzle.

While frustrated by their current failings, people may initially regard any changes as temporary, and are sometimes confident of a good, if not a complete, recovery. A gradual and painful process of adjustment may follow, when the person realizes the full extent of his/her residual disabilities and an appreciation that he/she will not after all recover former skills and life style. This process of recognition may result in a period of depression, characterized by 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, as they attempt to make sense of the condition. They may, for example, set themselves unrealistic targets, which may lead to repeated disappointment and despair.

Webb (1998: 543) highlights the complex nature of brain injury. The primary brain injury is often combined with secondary psychological difficulties. In particular, he reminds readers that 'the triumph of the body is poor compensation for the sequestration of the mind...' and that a lack of an obvious physical impairment results in those affected by acquired brain injury appearing normal to others. Webb (1998: 549) warns of the risk that '...those who are mind-impaired are consigned to the wastelands of social exclusion'.

Neurological diseases and injuries, including encephalitis, are therefore multi-faceted conditions with uncertain prognoses that have far-reaching consequences for those affected and their families.

Family/caregiver outcomes

The family of a person with encephalitis often struggles to offer support, while at the same time coming to terms with changes in their loved one. Webb (1998: 550) observed what he termed the 'bankruptcy of identity' (i.e. a loss of sense of self, or a loss of self-identity) that loved ones often bear witness to in respect of their brain-injured loved one. Family members provide practical, social and emotional support for their loved ones and this can have important implications for the carer's own life. Research by Man (2002: 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) concurred with such findings when they found that anxiety and depression were more likely to be present in those responsible for the care of a brain-injured relative.

Social outcomes

An understanding of how people cope and make sense of encephalitis is only beginning to be developed.

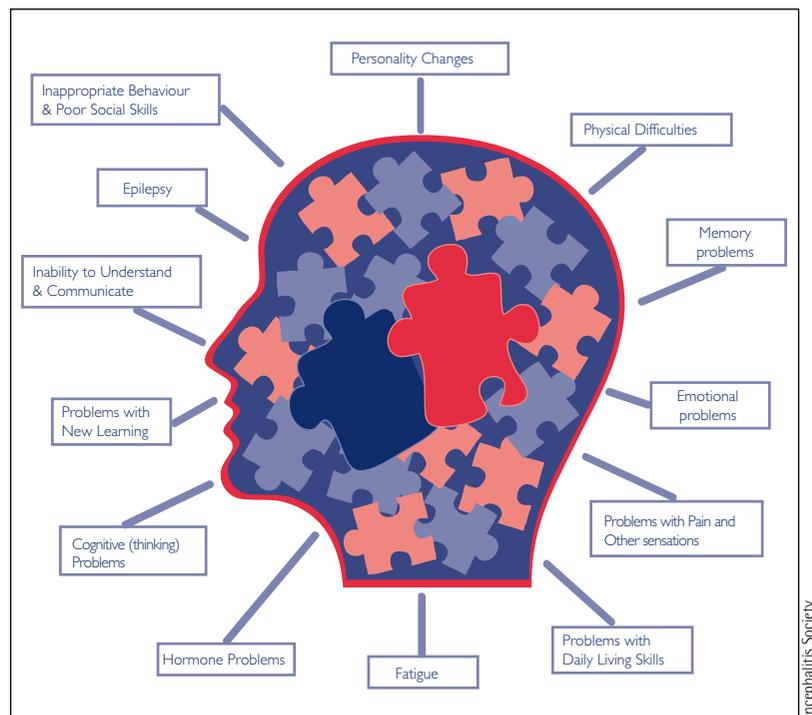


Figure 1. The consequences of encephalitis.

Much the same can be said for other neurological disorders or acquired brain injury. However, it would be fair to observe that certain types of neurological condition, such as traumatic brain injury and stroke, have received much more attention than encephalitis in recent years.

Encephalitis and other forms of neurological disease and injury restrict the lives of those people affected in a variety of ways. They disrupt social and family life, cause absences from work. Moreover, they have profound consequences for a person's psychological wellbeing, challenging the person's sense of self and affecting relationships with others. Macniven et al (2003: 528) documented the experience of Kate, who had acute disseminated encephalomyelitis, illustrating the impact of being wrenched out of one's reality:

Following her illness, Kate's relationships, employment, social existence and identity had all altered . . . her illness appeared to have changed everything.

Individuals may also face disadvantage and find themselves excluded from activities that the general population take for granted. They might encounter prejudice and negative attitudes, both socially and as they engage with peers, employers and financial institutions. This adds to their sense of social exclusion and discrimination. Webb (1998) reminds readers that the disabilities of a brain-injured person are not readily addressed by the use of technology, and that people with acquired brain injury 'have a greater propensity to be socially excluded' than those whose disability is physical.

Rehabilitation

Given the nature of the brain, the consequences of encephalitis can potentially be considerably greater than for an injury of any other organ. The brain, through the central and peripheral nervous systems, controls all of the systems which make up the body. This inevitably means that representatives from many disciplines are required to tackle the potential consequences of a brain injury. Rehabilitation needs to be provided in an interdisciplinary context, so that different aspects of impairment can be addressed by specialists. One approach to rehabilitation that embraces the complexity of acquired brain injury is the biopsychosocial model of rehabilitation (Williams and Evans, 2003).

Many people who survive encephalitis have some recollection of their previous lives. This can make their self-esteem fragile because they know the extent of their impairments. It is therefore critical that the care given to the person by each member of the rehabilitation team is consistent in providing a nurturing and supportive environment.

Neuropsychological assessment

A neuropsychological assessment is an important tool in planning appropriate care and rehabilitation for people affected by encephalitis. Neuropsychologists are trained in understanding how a person's emotions and cognition is related to the functioning of his/her brain. Assessment entails an appraisal of the brain's psychological, cognitive and behavioural functions. The assessment should highlight any primary problems (e.g. in concentration, memory, speed of thought), the nature of that difficulty and the best ways in which to address these problems.

A neuropsychological assessment should always be followed by neuropsychological rehabilitation. Subsequent assessments can help chart the process of change in symptoms over time. It is important to document improvement over time because care plans may require revision and the person's need for care services and resources may change and need re-assessment.

A lack of services and research

Little is known about the social consequences of encephalitis. Understanding is lacking, in particular, about how people affected make sense of their experiences, not only of the acute stage of the illness (much of which cannot usually be remembered) but also of any subsequent reduction in function, and the impact this has on their lives. The majority of the literature that currently exists on encephalitis is concerned with acute clinical and pharmacological management, life saving procedures, and psychological findings and debates. Little has been written about the quality of people's lives once they have been saved. It could be argued that quality of life has been relegated a secondary importance to the clinical treatment of the condition. It is important to be mindful therefore that such a bias may contribute to the ongoing medicalization of a person's illness experience.

Perhaps it is unsurprising then that Moorthi et al (1999: 140) observes that 'the literature on this subject is scant, probably due to the infrequent appearance of patients with encephalitis in inpatient rehabilitation programmes'. It is perhaps even more surprising, however, to learn that despite the severity of the disabling consequences of encephalitis (Moorthi et al, 1999: 140; Raschilas et al, 2002), there has been a historical lack of rehabilitative care for this group of people. It is therefore important that 'regardless of whether a specific cause is found, supportive care and early referral for rehabilitation maximise functional recovery' (Lewis and Glaser, 2005: 356).

It is important to question why there have been so few studies on the long-term or psychosocial outcomes of encephalitis and why people with encephalitis do not regularly receive referral for rehabilitation. One influential factor might be that, even though encephalitis is a notifiable disease, 97% of hospitalized cases go unreported (Davison et al, 2003). A further complication might be the dearth of neurologists in the UK. It has been reported in *The Lancet Neurology* (Anon, 2004) that there was the equivalent of 352 full-time consultant neurologists in the UK: one per 170 000 people. This figure is described as 'lagging far behind other countries in Europe'; astonishing especially when it is considered that the number of people with a neurological disorder in the UK is estimated at 10 million (Neurological Alliance, 2003).

An evaluation of Encephalitis Society members' self-reported experiences of neurological services in the UK ($n=339$) found that many people had had no access to the care of a neurologist (37%), and some of these had not seen any kind of specialist (Easton, 2005). Of those who did see a neurologist, fewer than half saw one within 24 hours of admission to hospital, and over one third had to wait more than 3 days. At least half of the respondents to the survey (56%) reported not being cared for on a neurology ward. Of those who were, fewer than a quarter were transferred within 24 hours of admission.

Such findings go some way toward explaining why encephalitis receives less focus than other, more common neurological conditions, or conditions that are less common but which may have a higher profile or public focus, such as motor neurone disease, multiple sclerosis and brain tumours. The result is that people who have been affected by encephalitis obtain less specialized treatment and therefore have fewer opportunities to come to terms with the consequences of their brain injury. Moreover, encephalitis is not a public health priority and little research is undertaken to investigate its consequences and social impact. The treatments available for viral diseases are limited and aciclovir has been a generic drug for many years. Moreover, the pharmaceutical industry may find little incentive to invest in research to develop new treatments for a disease such as encephalitis which affects a relatively small group of the population.

The encephalitis patient might therefore be considered an orphan of the health system. Encephalitis patients are in a minority and because of this there is a lack of

awareness not only among the general population but also among those best placed to help them: the health and social care professions.

Conclusions

Encephalitis is a relatively rare condition that can result in acquired brain injury. The consequences of encephalitis can be greater than for some other types of brain injury and can vary from a slight change in mental ability to profound disability, physical, mental or both. Among a plethora of cognitive, physical, behavioural or psychosocial difficulties, emotional problems are common and need understanding and sensitive handling. A small percentage of those affected by encephalitis will need to remain in residential care for the rest of their lives. A neuropsychological assessment is an important tool in planning appropriate care and rehabilitation.

Sadly, access to specialist rehabilitation, although patchy, is still less for people affected by encephalitis, than for some other types of acquired brain injury. Greater awareness of encephalitis and its consequences among the general public and among health, education and social care professionals will help reduce the number of people affected by encephalitis who fall through the gaps in the provision of services.

Conflict of interest: None declared

- Anon (2004) Wanted: Better neurology service. *Lancet Neurol* **3**: 507
- Aygun AD, Kabakus N, Celik I, Turgut M, Yoldas T, Gok U, Guler R (2001) Long-term neurological outcome of acute encephalitis. *Journal of Tropical Pediatrics* **47**: 243–7
- Beghi E, Nicolosi A, Kurland L, Mulder D, Hauser A, Shuster L (1984) Encephalitis and aseptic meningitis, Olmstead County, Minnesota, 1950–1981: I. Epidemiology. *Ann Neurol* **16**: 283–94
- Booss J, Esiri M (2003) *Viral Encephalitis in Humans*. ASM Press, Washington DC
- Clark L, Barriball LK (2006) Inclusion in the mainstream: Challenges in the care of people with intellectual impairment. *British Journal of Neuroscience Nursing* **2**(5): 227–31
- Davison KL, Crowcroft NS, Ramsey ME, Brown DWG, Andrews NJ (2003) Viral encephalitis in England, 1989–1998: What did we miss? *Emerging Infectious Diseases* **9**(2): 234–40
- Dowell E, Easton A, Solomon T (2001) *The Consequences of Encephalitis*. The Encephalitis Society, London
- Easton A (2005) Encephalitis patient experiences of neurological services in the UK. World Brain Injury Congress, Melbourne, Australia, 6–8 May
- The Encephalitis Society (2003) Encephalitis (general leaflet). The Encephalitis Society, Malton
- Glaser CA, Gilliam S, Schmurr D, Forghani B, Honormand S, Khetsuriani N, Fischer M et al (2003) In search of encephalitis etiologies: Diagnostic challenges in the California Encephalitis Project, 1998–2000. *Clin Infect Dis* **36**: 731–43
- Khetsuriani N, Holman RC, Anderson LJ (2002) Burden of encephalitis-associated hospitalizations in the United States, 1988–1997. *Clin Infect Dis* **35**: 175–182
- Lewis P, Glaser CA (2005) Encephalitis. *Pediatrics in Review* **26**(10): 347–57
- McGrath N, Anderson NE, Crosson MC, Powell KF (1997) Herpes simplex encephalitis treated with acyclovir: Diagnosis and long-term outcome. *J Neurol Neurosurg Psychiatry* **63**: 321–6
- Macniven JA, Poz R, Bainbridge K, Gracey F, Wilson BA (2003) Emotional adjustment following cognitive recovery from 'persistent vegetative state': Psychological and personal perspectives. *Brain Injury* **17**(6): 525–33

KEY POINTS

- Encephalitis is an inflammation of the brain usually caused by a viral infection
- Some loss of brain function is a probable outcome of encephalitis
- The neurological, psychological and social consequences of encephalitis are often poorly understood
- Rehabilitation of people affected by encephalitis may benefit from interdisciplinary and biopsychosocial approaches
- Neuropsychological assessment can be a useful tool in recovery

Man DWK (2002) Family caregivers' reactions and coping for persons with brain injury." *Brain Injury* **16**(12): 1025–37

Moorthi S, Schneider WN, Dombovy ML (1999) Rehabilitation Outcomes in Encephalitis - a retrospective study 1990–1997. *Brain Injury* **13**(2): 139–46

The Neurological Alliance (2003) Neuro numbers: A brief review of the numbers of people in the UK with a neurological condition. The Neurological Alliance, London

Ponsford J, Olver J, Ponsford M, Nelms R (2003) Long-term adjustment of families following traumatic brain injury where comprehensive rehabilitation has been provided. *Brain Injury* **17**(6): 453–68

Raschilas F, Wolff M, Delatour F, Chauffaut C, De Broucker T, Chevret S, Lebon P et al (2002) Outcome of and prognostic factors for herpes simplex encephalitis in adult patients: Results of a multicenter study. *Communicable Infectious Diseases* **35**(1): 254–60

Schott J (2006) Limbic encephalitis: A clinician's guide. *Practical Neurology* **6**: 143–53

Webb D (1998) A 'revenge' on modern times: Notes on traumatic brain injury." *Sociology* **32**(3): 541–55

Whitley RJ, Gnann JW (2002) Viral encephalitis: Familiar infections and emerging pathogens." *The Lancet* **359**: 507–13

Williams WH, Evans JJ (2003) Bio-psychosocial approaches for mood and behaviour disorders in neuro-rehabilitation: An overview. *Neuropsychological Rehabilitation* **13**(1–2): 1–12

Further information

The Encephalitis Society

www.encephalitis.info

7B Saville Street, Malton, North Yorkshire YO17 7LL

The Encephalitis Society is a national charity and the only organization of its kind in the world providing dedicated support to people affected by the illness and their families. Support is provided by telephone, letter, email and personal contact in the UK and the Republic of Ireland, and by email to people across the globe. The Society has a comprehensive resource of information available in both printed and electronic form, and can also provide training to health, social care and education professionals on request.

Helpline

Email: support@encephalitis.info

Tel: 01653 699 599

Training for Professionals

Email: training@encephalitis.info