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Managing severe epilepsy in the community

Stephen W. Brown

The term 'epilepsy' refers to a tendency to have recurrent seizures. Epileptic seizures arise from an imbalance of excitatory and inhibitory cerebral neurotransmitters associated with sudden, paroxysmal, synchronous and repetitive discharges of neurones. All normal brains have the capability to have seizures. Epileptic seizures arise where there is lowering of the naturally occurring seizure threshold. Excitotoxins such as glutamate released in the brain during seizures can cause irreversible cell damage. Young children are especially vulnerable. Prolonged seizures (greater than 30 minutes) in children correlate with subsequent learning disability.

People with learning disabilities are at particularly high risk of developing seizure disorders. Seizures are often difficult to treat in this population and present special problems in diagnosis, investigation and overall management. Neurological services are not usually well set up to deal adequately with these sorts of problems. Seizures, and their treatment, may also affect behaviour, emotions and cognitive functioning. It therefore behoves psychiatrists, especially those who treat people with learning disabilities, to be familiar with the diagnosis and management of epilepsy.

Epidemiology

Epilepsy is the most common serious neurological condition. In North America and in Europe the lifetime risk of developing epilepsy is about one in 30. The prevalence rate in the general population is about 0.6–0.7%. The lifetime risk and the point prevalence are different for several reasons. Epilepsy is not always a lifelong condition, and the onset of the various forms of epilepsy occurs at different ages. Also, epilepsy carries a significant mortality.

In the population with learning disabilities the prevalence of epilepsy is much higher than in the general population. Among people with learning disabilities who are in touch with specialist services about 30% have epilepsy. In institutionalised settings this figure may approach 50%. Even in those with mild learning disabilities the prevalence rate is about 6%, 10 times greater than in the general population (Hopkins & Shorvon, 1995; Cassidy & Corbett, 1997).

Health loss in epilepsy may be related to the following factors:

(a) There is an excess mortality. Tonic–clonic seizures may be associated with sudden death, which may be a result of central apnoea or cardiac dysrhythmia (Nashef & Brown, 1996). In people with severe refractory epilepsy about one in 100 may die each year in this way. Death may also occur accidentally as a result of a seizure, for example by drowning. Epilepsy also carries an excess risk of suicide.

(b) Epileptic seizures can also directly affect health by causing injuries. Recurrent head injuries may have deleterious long-term effects on cognitive functioning.

(c) The treatment of epilepsy may have adverse effects on health. Anti-epileptic drugs can affect cognition, mood and behaviour as well as causing the other neurological symptoms, and may affect other systems causing problems ranging from cosmetic side-effects to teratogenesis.

(d) A diagnosis of epilepsy often carries a social stigma, which may affect normal psychological development. There may also be restrictions on activities and job opportunities.

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Patterns of severe epilepsy

About 50 different types of epileptic seizure are known. One person may have more than one seizure type. A precise description of the seizure types, taken together with other relevant information such as age of onset and electroecephalogram (EEG) findings, enables a diagnosis to be made at the level of an epilepsy syndrome. The seizure type itself is a clinical feature, rather than a diagnosis (International League Against Epilepsy, 1989, 1993).

Seizure types

By international agreement seizures are classified into those that begin on one side of the brain (partial), and those in which both sides of the brain are involved at the onset (known as generalised).

Partial seizures are further subdivided into 'simple partial seizures', where consciousness is retained, and 'complex partial seizures' in which consciousness is impaired. Partial seizures may evolve to become secondarily generalised. Complex partial seizures include events that have previously been called psychomotor seizures. Simple partial seizures include Jacksonian motor events, as well as sensory phenomena including brief premonitory feelings that some people get before a generalised seizure, which are known as 'auras'.

Generalised seizure types include tonic–clonic (grand mal) seizures, absence seizures and akinetic seizures or drop attacks. It is important to note that tonic–clonic seizures without any history of aura are nevertheless frequently secondary generalised in type, because generalisation may be rapid, or because there may be amnesia for the period just before the tonic–clonic seizure.

In a tonic–clonic seizure there may occasionally be a preceding prodromal mood change lasting several hours, sometimes more noticeable to the patient's family or carers than to the patient. This is a different phenomenon to an aura.

The tonic–clonic seizure itself starts with a phase of stiffening which lasts for a few seconds (the tonic phase). Sometimes this may begin in one limb or side of the body, but often there is no obvious laterality. The tonic phase proceeds to a phase of convulsing which may last for minutes and which gradually subsides. This is then typically followed by a period of post-ictal confusion and/or sleep, which may last minutes or hours. The tongue may be bitten in a tonic–clonic seizure, and there may be incontinence of urine. Occasionally, people may stop breathing and die in a seizure.

Epileptic syndromes

Seizure types, together with other data such as age of onset and EEG findings, constitute epileptic syndromes. Like seizure types, these are divided into two major groups depending on whether the seizures start on one side or both sides of the brain. These are referred to as 'localisation-related epilepsies' (such as temporal lobe epilepsy) and 'generalised epilepsies' respectively.

The generalised epilepsies include idiopathic syndromes such as childhood absence epilepsy and juvenile myoclonic epilepsy. This group, depending on the precise syndrome, tends to be characterised by absence seizures, myoclonic seizures and tonic–clonic seizures. There may be a genetic component, and they tend not to be associated with other neurological problems.

The other main grouping of generalised epileptic syndromes is the 'cryptogenic' or 'symptomatic' group. These are of special interest to learning disability psychiatrists because they tend to be associated with cerebral insult in early life, or a neurodevelopmental disorder, and they include West syndrome (infantile spasms), which may be associated with tuberous sclerosis and the Lennox–Gastaut syndrome.

Syndromes associated with learning disability

West syndrome (infantile spasms)

This condition has been recognised since West described it in his own child in 1841. The age of onset is usually between four and seven months and is always before the age of one year. The spasms are generalised brief myoclonic contractions of the neck, trunk and extremities, which may occur both as single seizures and in series. There is a characteristic inter-ictal EEG pattern called hypsarrhythmia. In many cases the spasms may disappear within one year after the onset. Further cognitive development may be normal if the spasms are controlled, which may happen in 10–30% of cases. However, severe learning disability, often with associated pervasive developmental disorder, is the more frequent outcome. Other seizure types may occur later and in some cases the condition merges into the Lennox–Gastaut syndrome.

About 80% of cases are symptomatic and the underlying aetiologies may be tuberous sclerosis or one of a number of other conditions including perinatal asphyxia, cortical dysplasia and neuro-

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fibromatosis. The prognosis is mainly influenced by the aetiology.

Lennox–Gastaut syndrome

This is one of the most common malignant epilepsies beginning in childhood and is seen in the population with learning disabilities. The age of onset is usually between one and five years. Many seizure types may occur, but the most typical are axial tonic seizures, atonic seizures and atypical absences (these latter are characterised by slow spike–wave on the EEG). Tonic–clonic and complex partial seizures may also occur. Non-convulsive status epilepticus is common. The EEG shows generalised slow spike–wave with other epileptiform features especially polyspike, against a slowed background rhythm. The onset is sometimes preceded by a neurological illness. In such cases there is developmental arrest and the patient may have severe learning problems and a loss of skills. A very small number of patients fall within the average range of abilities. The seizures may attenuate in adult life.

Mesial temporal lobe epilepsy

This is probably the most common type of epilepsy, and is found in people with and without learning disabilities. It is characterised by: (a) simple partial seizures (most typically a rising sensation in the abdomen, but other autonomic or psychic symptoms such as fear, or olfactory and auditory hallucinations may occur); (b) complex partial seizures (often beginning with motor arrest followed by an oro-alimentary automatism); and (c) secondarily generalised seizures, or combinations of these. The complex partial seizures seen in this syndrome may be marked by limb automatisms ipsilateral to the side of onset with contralateral dystonic posturing. An inter-ictal scalp EEG may show no abnormality, or may show asymmetry of background activity, or may show focal epileptiform discharges. There may be a history of febrile convulsions in infancy. Discrete memory deficits may be demonstrated on formal testing. The age of onset is frequently in childhood or adolescence. The pathological lesion associated with this syndrome is mesial temporal sclerosis, but the precise aetiology remains unclear.

Frontal lobe epilepsy

Frontal lobe epilepsy is also characterised by simple partial, complex partial, secondarily generalised seizures or combinations of these. The seizures often occur several times a day, are of short duration and secondary generalisation is very rapid. They frequently occur during sleep. The seizures may contain prominent motor manifestations, which are tonic or postural, and complex gestural automatisms are frequent at the onset. The seizures of frontal lobe epilepsy are often mistaken for non-epileptic events. Recently a specific genetic condition, autosomal dominant nocturnal frontal lobe epilepsy, has been described (Scheffer *et al*, 1995).

Landau–Kleffner syndrome (acquired epileptic aphasia)

In this condition, which usually begins between the ages of 3–8 years, an acquired aphasia is associated with multi-focal spike, or spike and wave discharges. Epileptic seizures occur in about two-thirds of cases. There is a receptive and expressive aphasia with an associated behavioural disturbance, which may have features of attention deficit hyperactivity disorder, while severe cases may resemble a disintegrative psychosis. A recent observation has been that the onset of the syndrome may be associated with electrical status epilepticus in sleep. This latter condition may also occur without aphasia, and it seems that Landau–Kleffner syndrome may represent a subtype of a broader continuum of disorder. The seizures and EEG abnormalities usually remit by the age of 15 years, but the person is typically left with a degree of learning disability (Smith, 1997).

Behavioural changes and psychiatric associations

Epilepsy is a risk factor for increased psychiatric morbidity (Rutter *et al*, 1970). One recent population-based study in Sweden found a rate of psychiatric diagnosis as high as 59% in children with a combination of learning disabilities and epilepsy. Many of these problems had been undiagnosed despite parental concern (Steffenburg *et al*, 1996). Fenwick (1995) has suggested that in the general adult population 30–50% of people with epilepsy have an identifiable psychiatric problem, albeit often minor. Where comparative studies have been made, it seems that the rate of psychiatric morbidity in people with learning disabilities who have epilepsy is not significantly different to that of the rest of the population who have epilepsy (Espie *et al*, 1989). However, Deb & Hunter (1991*a*) have suggested that there is a subgroup of people with mild learning disabilities who have refractory epilepsy with generalised EEG epileptiform discharges, and who have an increased rate of behaviour disorder. In a

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related study (Deb & Hunter, 1991*b*) the authors described a slight increase in the expected rate of schizophrenia and delusional disorder in the population with epilepsy and learning disabilities. The overall rate of psychiatric diagnosis was 25%.

Special clinical problems

Non-convulsive status epilepticus

Status epilepticus is defined as a fixed epileptic state lasting 30 minutes or longer. It may be convulsive, involving repeated tonic–clonic seizures (which is a well-known medical emergency), or non-convulsive, either due to continuous absence seizures or repeated complex partial seizures.

In absence status there is clouding of consciousness which varies over hours or days. The features can vary from stupor to subtle behavioural changes. Although it is common in children and young people with learning disabilities, absence status may be the presenting feature of epilepsy in the elderly, and is therefore another item to add to the list of causes of confusional states in older people.

In complex partial status, repeated stereotyped behaviours are usually seen against a background confusional state. The stereotyped behaviours represent the automatism of the seizure. Psychotic features are seen with or without clouding of consciousness. The repetitive nature of the behaviour becomes apparent with careful observation, but may be missed on casual examination. Complex partial status may also present as a psychotic state without obvious repeated stereotypies, especially if it is of frontal or other non-temporal origin. In either case the ictal causation is often missed, and a misdiagnosis of schizophrenic or schizoaffective psychosis is made.

Typically, an EEG will confirm the diagnosis of non-convulsive status epilepticus, so this investigation should be available to people with learning disabilities and it should be possible to obtain it when it is needed.

Non-convulsive status is common in Lennox–Gastaut syndrome, and may last for weeks at a time. Unresponsiveness, disorientation, dribbling and a need for help with basic functions like eating, dressing and washing, may all be consequences. Unfortunately, in the population with learning disabilities carers may attribute such features to the person's constitution, and the diagnosis is missed. A high index of suspicion is therefore required (for a review of non-convulsive status epilepticus in people with learning disability, see Staufenberg & Brown, 1994).

Post-ictal psychosis

This is an interesting self-limiting condition, the prevalence of which is unknown. It has been properly described in the modern literature by Logsdail & Toone (1988). Typically, a cluster of tonic–clonic seizures is followed by a latent interval lasting 1–6 days, and is followed by a florid psychosis which usually subsides over the following days or weeks. Males are said to be more affected than females, and the condition may occasionally occur after an isolated tonic–clonic seizure, or after a series of complex partial seizures. One study suggested that post-ictal psychosis may be characterised more by grandiose or religious delusions and mystical experiences and therefore differs from other types of psychosis in its phenomenology (Kanemoto *et al*, 1996).

Paricta l psychosis

Strictly speaking, post-ictal psychosis has an onset after a seizure or cluster of seizures. Less commonly, psychotic symptoms may develop in parallel to an increase in seizure frequency, which if rapid may be confused with post-ictal psychosis. Here, treatment must be primarily directed to bring the epilepsy under control. This latter phenomenon has been called paricta l psychosis (Trimble & Schmitz, 1997).

Inter-ictal psychosis

It remains controversial whether the rate of psychoses not directly related to ictal events is increased in people with epilepsy compared with the rest of the population. It does seem, however, that there is an increased rate in people who attend specialist centres for epilepsy. This includes people with learning disabilities and severe refractory epilepsy. Risk factors for developing inter-ictal psychosis have been summarised by Trimble (1991). They include a slight bias to females, having temporal lobe epilepsy, an age of onset in early adolescence, a mean gap between the onset of epilepsy and onset of the psychosis of 14 years and a left-sided focus. Seizure frequency is often reported to be lower in patients who have a chronic psychosis than in those who do not. The reason for this is unclear. A small group of patients has shown a phenomenon where psychotic features occur if seizures become much less frequent or disappear. An increase in seizure frequency is accompanied by a decrease in psychotic features. This is known as an alternating psychosis, and the phenomenon seen in the EEG, where the epileptiform features may

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disappear when the psychosis appears, is called paradoxical normalisation. It may, of course, be a consequence of changing drug treatment for the epilepsy, although in some cases it seems to occur spontaneously.

Non-epileptic seizures

It is a frequently reported observation that a high proportion of people who have a diagnosis of epilepsy actually have seizures which are not epileptic. This has been estimated as accounting for between 20 and 30% of patients attending specialist clinics. Non-epileptic seizures may occur in people who have epilepsy or may be found in the absence of epilepsy. Non-epileptic seizures may have mainly physiological causes or mainly psychological causes. The mainly physiological causes include syncope, migraine and parasomnias. The mainly psychological causes include hyperventilation or panic disorder and a group of conditions collectively referred to as non-epileptic attack disorder.

The latter includes non-epileptic seizures which have in the past been referred to as hysterical seizures, psychogenic seizures or pseudoseizures. Non-epileptic attack disoder is more common in females. Concomitant psychiatric disorders include affective disorder, dissociative states and post-traumatic stress disorder. Some special features, which may be found in children, include unresponsiveness and generalised violent and uncoordinated movements. The general features of non-epileptic attack disorder which may help distinguish these events from true epileptic seizures include longer ictal duration, less stereotypy, asynchronous limb movements, alternating head movements and pelvic thrusting. The latter symptom is said to be more common in young females who may have a history of sexual abuse. In these cases, post-traumatic stress disorder may manifest as a seizure disorder. The person experiences flashbacks and enters a dissociative state, which bears a superficial resemblance to an epileptic seizure. Typically, the patient does not report memory for the seizure event afterwards. These types of seizures may be reported as occurring during sleep, but if the patient is monitored, it can be seen that the person awakes from sleep before the onset of the seizure.

Other observations regarding non-epileptic attack disorder include the presence of an increased rate of soft neurological signs and a history of previous brain injury (Scheepers *et al*, 1994). These may arise in families where there is less cohesion than in the normal population, and less family emphasis on ethical issues and values (Moore *et al*, 1994).

Finally, there has been much interest in the use of post-ictal prolactin estimation as a method of distinguishing truly epilepsy from non-epileptic attacks (Collins *et al*, 1983; Anzola, 1993). To make use of this technique it is necessary to ensure that the patient has no other reason to have a raised prolactin level. The baseline reading must be taken at a time when the patient has not recently had a seizure. As a rule of thumb, a rise in prolactin level to three times over the baseline occurring 20 minutes or so after the onset of a seizure is highly suggestive of true epilepsy. This is because the anterior pituitary discharges during the seizure. Failure to demonstrate a prolactin rise does not of itself indicate a non-epileptic event.

Management issues

Diagnosis and investigations

Epilepsy is a clinical diagnosis, based on adequate history taking. Even in people who do not have a learning disability it is essential to obtain an eyewitness account of seizures. Investigations, such as EEGs, are useful to clarify the nature of the epileptic syndrome and to pursue aetiology. The EEG is mainly used to help define the epileptic syndrome. An inter-ictal EEG may be normal; for example, this is often the case in temporal lobe epilepsy, while abnormal EEGs are often reported in people with learning disabilities who do not have epilepsy. Intensive monitoring of the EEG is useful to help distinguish some non-epileptic events, but this sort of investigation needs to be carried out by experienced teams and is not necessary in the majority of cases.

It is considered good practice to try and seek a cause for a seizure disorder, especially where the seizures are of partial onset. People with localisation-related epilepsies should therefore be referred for neuroimaging wherever possible. The preferred imaging investigation is magnetic resonance scanning, although computerised tomography scanning is more widely available. It is also good practice in the management of learning disabilities to screen for metabolic and genetic aetiologies (Jenkins & Brown, 1992; Brown *et al*, 1993).

People with learning disabilities have as much right as any other members of the population to have access to investigations and scanning procedures. In fact, it could be argued that because of their special needs and the difficulties in treating their epilepsies,

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they should be given some degree of priority. It is, therefore, important that physicians who are responsible for treating people with learning disabilities are able to act as advocates in ensuring that this is the case.

Treating epilepsy

There are four key points in treating epilepsy. The single most important factor influencing the quality of life in people with epilepsy is whether seizures continue to occur despite treatment (Jacoby *et al*, 1996). Seizures are most directly associated with mortality and morbidity. It is inappropriate to regard a person's epilepsy as well controlled if seizures still occur. In people with learning disabilities, epilepsy can be very difficult to control.

Proper attention should be given to recognising side-effects of treatment. People with learning disabilities may have problems with communication and will not necessarily volunteer information about mood changes or sedation. Some neurological signs related to drug intoxication may be missed unless the physician makes a point of checking for them. However, one of the aims of treating epilepsy is to use a regime that minimises adverse effects of treatment.

Ideally, the patient should be free from adverse effects on everyday life as a consequence of being a patient or having epilepsy. This means organising the services for the convenience of the service user, avoiding unnecessary investigations and educating carers about the condition so that unnecessary restrictions on lifestyle are not made, while appropriate risk management can be carried out.

Patients and their carers are part of the team managing the epilepsy and should be able to play a full part in arriving at decisions about treatment and other aspects of management.

Treatments available for epilepsy include anti-epileptic drugs, surgery, dietary treatments, behavioural treatments, environmental manipulation and complementary medicine. For most people the mainstay of treatment will be anti-epileptic drugs, but the other aspects should not be overlooked.

Anti-epileptic drugs

The reason that the diagnosis of epilepsy has to be made at the level of an epileptic syndrome is that this will influence the choice of drug treatment. For example, carbamazepine can make some of the seizures in generalised epilepsy syndromes worse and should therefore be avoided.

Principles of prescribing

The general principles of prescribing anti-epileptic drugs in this population are to choose a drug according to the epileptic syndrome, and to start with monotherapy in a low dose. This should be escalated in small increments to a therapeutically effective level. The case for rational polytherapy has still to be proven. If the first drug is ineffective, another drug may be chosen and introduced alongside the first and built up to a therapeutic level. If there is a positive response, then the original drug should be slowly withdrawn.

Choice of anti-epileptic drug

There are, roughly speaking, three generations of anti-epileptic drugs in current use. Those in the first generation were mainly licensed before the Second World War. They include phenobarbitone and phenytoin. Few, if any, serious specialists in epilepsy in the UK today would recommend these as a first-line treatment for epilepsy, and especially not for people with learning disabilities. These drugs may sometimes be helpful in individual cases, but they all have potentially adverse effects on mood and behaviour. Phenytoin has the added disadvantage of eccentric pharmacokinetics and a range of possible long-term physical side-effects. The non-linear relationship between the dose of phenytoin and its serum level, and the wide range of adverse reactions makes a fascinating study for the clinical pharmacologist, but should serve as a warning to the prescriber.

The second generation, carbamazepine and valproate, licensed in the 1960s and early 1970s, are usually regarded as standard first-line anti-epileptic drugs today. Compared with the earlier drugs, they are said to have better side-effect profiles. Valproate can cause cosmetic side-effects such as weight gain and hair loss. Both drugs are teratogenic, and can be associated with neural tube defects, and this is especially true of valproate. Valproate can also be associated in the population with learning disabilities with hyperactivity and tremor, and carbamazepine is sometimes found to be a sedative. They have different indications. Valproate is a broad spectrum anti-epileptic drug whereas carbamazepine is really only effective in localisation-related epilepsies. Further, carbamazepine may precipitate absence seizures and may make myoclonus worse. Both drugs are available in controlled release preparations, which ensure continuity of formulation and allow a smaller number of doses per day to be taken with good effect. The controlled

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release preparation of carbamazepine in particular helps to minimise acute side-effects such as double vision. Valproate may be associated with hyperammonaemia, which in turn may be aggravated by concomitant carnitine deficiency.

Drugs in the third generation have all achieved their licence in the UK since 1989. There are currently four new anti-epileptic drugs in this group and a fifth is expected in 1998. Each has a different mode of action and its own special practice points. It has been suggested that these newer generation drugs may be of special value not only because of the potential for greater efficacy, but also because they may provide better safety profiles and fewer interactions with other drugs (Mattson, 1996). Table 1 summarises some of the key points of these new drugs.

Which drug for which syndrome?

Generally speaking, anti-epileptic drugs may be regarded as either 'broad' or 'narrow' spectrum in their actions. The latter tend only to be effective in localisation-related epilepsies, and include carbamazepine, gabapentin, tiagabine and vigabatrin. The broad spectrum drugs, which may be used in both generalised and localisation-related syndromes, include valproate, lamotrigine, topiramate and the benzodiazepines.

Tiagabine is likely to be launched in the UK 1998 or 1999. Because it is a drug which acts on γ-aminobutyric acid systems there is at least a theoretical possibility that it may be associated with visual field defects.

People with learning disabilities are at much higher risk of developing cryptogenic and symptomatic generalised epilepsy syndromes, such as Lennox–Gastaut syndrome, than the rest of the population. There is an emerging view that lamotrigine, with its broad spectrum of activity and lack of sedative side-effects should be the drug of choice in this group. It does currently have a licence for Lennox–Gastaut syndrome. As far as the localisation-related epilepsies are concerned, the traditional first-line treatment is carbamazepine. This is usually well tolerated although it can be sedative and can depress white cells. Some people also experience minor skin problems. Gabapentin, currently licensed as an add-on treatment, is likely to obtain a monotherapy licence in 1998 or 1999, and represents an alternative, which is usually well tolerated.

Community management

With the exception of phenytoin, it is not necessary routinely to measure serum drug levels. If a patient is taking a combination of medications, and there are clinical signs of toxicity, such as cerebellar signs, then a blood test may be helpful in deciding which drug is causing the problem. Also, drug levels can be tested as compliance checks.

It is helpful to know how many seizures the patient is having. Proper treatment diary cards are available from several of the drug companies and some clinics devise their own. Patients or their carers should record the frequency and timing of the seizures so that progress should be monitored. Patient medical notes should contain a record of current seizure frequency each time the patient is seen. In a busy clinic it is easy to overlook recording these details, but it is important.

There are several other factors, which can act as measures of efficacy of intervention. Crude seizure counts may not be sufficient in this respect. A change of treatment may have an effect on day centre attendance, on the requirement for rectal diazepam, or on attendances at the general practitioner's surgery or at the accident and emergency department.

Behavioural consequences of anti-epileptic drug treatment

Changes in mental state, which vary with seizure rate and type, are recognised phenomena in the natural history of complex epilepsy. They may be related to anti-epileptic drugs in a number of ways.

Any anti-epileptic drug may be associated with a paradoxical normalisation psychosis (also called 'forced normalisation') where sudden decrease in seizures (and typically the normalisation of the EEG) may precipitate symptoms of psychosis. This is bound to happen with any agent that is effective at stopping seizures. Its occurrence merely indicates that the anti-epileptic drug is effective.

Any anti-epileptic drug may cause a decrease in frequency of major seizure types, such as tonic–clonic seizures, with a change in pattern to having more minor seizures such as complex partials. These minor seizure types may present as behaviour changes, and may even present as complex partial status epilepticus mimicking a psychotic illness (Brown, 1993). These phenomena may follow any effective anti-epileptic drug treatment, and might respond to increasing the dose of medication. They do not represent side-effects of drug treatment as such.

Any anti-epileptic drug may cause a decrease in seizure frequency such that there is a persistence of prodromal mood or behavioural changes which usually precede seizures, and which usually cease when the seizure occurs. If a prodrome becomes prolonged this may present as mood or behaviour change. Such a change is only attributable to the

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Table 1. New anti-epileptic drugs

	Vigabatrin (UK licence 1989)	Lamotrigine (UK licence 1991)	Gabepentin (UK licence 1993)	Topiramate (UK licence 1995)	Tiagabine (UK launch 1998)
Mode of action	Suicidal inhibitor γ-aminobutyric acid transaminase	Inhibits paroxysmal glutamate release blocking rapid repetitive firing in sodium channels	Acts on alpha-2-delta subunit of N-methyl-D-aspartate receptor mediating calcium channel	Acts on both excitatory and inhibitory systems	γ-aminobutyric acid reuptake inhibitor
Indications	Infantile spasms, especially due to tuberous sclerosis, partial onset seizures. *NB. May make absences and myoclonus worse*	Broad spectrum, now also licensed in Lennox–Gastaut syndrome	Partial onset seizures	Licensed for partial onset, but recent studies suggest broad spectrum of use	Partial onset seizures
Important side-effects	Psychosis Behaviour change Visual field defect	Potentially allergenic (rash or extremely rarely Stevens–Johnson syndrome) May be associated with increased alertness, especially in the presence of valproate Double vision/ataxia may occur in presence of carbamazepine (and responds to lowering the dose of carbamazepine)	Usually well tolerated: one of the most commonly reported side-effects is increase in seizures, but it is not clear whether this is a particular effect of gabapentin or not, as all anti-epileptic drugs may do this	Cognitive slowing (may improve with continued use) Behavioural changes Possible risk of renal calculus (noted before launch, but not a problem since) Mild hepatic enzyme inducer, so potential interaction with oral contraceptives	Dizziness Tremor Cognitive problems (in high doses) Anecdotal reports of behavioural changes
Practice points	Minimise potential behavioural changes by using lower dose escalation than data sheet for population with learning disabilities, e.g. 250–500 mg/day in adults, increasing by 250–500 mg/day every 1–2 weeks up to a final dose range 2000–4000 mg/day *NB Current studies ongoing into significance of reported visual field defects: until this is clarified exercise discretion in initiating new prescriptions It is recommended that Humphrey visual field testing is carried out at six-monthly intervals in those patients able to cooperate*	Serum level is greatly increased by concomitant valproate so use lower dose regime if valproate present Use slow dose escalation to minimise risk of rash, e.g. 25 mg every other day in adults also taking valproate, doubling every 2 weeks to 100 mg/day, thereafter build up in 25–50 mg increments. Double this regime in the absence of valproate Some people tolerate very high doses (e.g. 800 or 1000 mg/day in absence of valproate) with good effect in epilepsy Licensed in monotherapy	Although generally well tolerated, it is still advisable to use a slower dose escalation in this population than that in the data sheet, e.g. in adults 300–400 mg/day for 1 week, increasing by 300–400 mg/day every week to 1800–2400 mg/day Some patients may benefit from (and tolerate) higher doses than data sheet, e.g. up to 4800 mg/day The contents of the capsules may be taken dissolved in blackcurrant cordial for people who cannot tolerate swallowing capsules Twice daily dosing mostly OK despite the short half-life	In the population with learning disabilities best to use slow dose escalation, e.g. in adults 25 mg every other day for 1–2 weeks, then 25 mg/day for 1–2 then increase by 25 mg every 1–2 weeks up to 200 mg/day. Careful watch for behavioural/cognitive problems Keep fluid intake up to minimise renal calculus risk	Short half-life (4–9 hours); 3 or 4 times daily dosing is more effective than twice daily May need higher doses in presence of enzyme inducing drugs, e.g. carbamazepine Still unclear whether special dose regime is needed in learning disabilities Still unclear whether visual field defects may be a problem

anti-epileptic drug in the sense that it is a consequence of incomplete treatment of the epilepsy. It should not be regarded as a side-effect of a drug in the usual meaning of this term.

Any anti-epileptic drug may have a paradoxical effect of increasing seizure frequency. This may theoretically result in (a) complex partial status or (b) post-ictal psychosis. In both cases the side-effect is the increase in seizure frequency, and the psychotic phenomena are consequences of the seizures, not the drugs.

Pre-ictal, ictal, post-ictal and inter-ictal psychotic phenomena, including paradoxical normalisation and post-ictal psychosis, may occur in people with epilepsy and be completely unrelated to changes in drug treatment.

An anti-epileptic drug might precipitate an inter-ictal psychosis unrelated to seizures, and if so it presumably could do this in people without epilepsy too. It is fairly widely accepted that vigabatrin can do this, and there is anecdotal evidence that topiramate might, but there is no real evidence that any of the other anti-epileptic drugs do.

A patient may have had many minor seizures per day for many years causing the appearance of chronic lethargy, often with psychomotor retardation and sometimes even feeding difficulties (especially in people with learning disabilities). Carers and family may regard this as part of the person's normal personality. Treatment with a new anti-epileptic drug, which successfully treats the epilepsy, means that the patient is no longer lethargic and slow, and this may manifest as a personality change. The appearance of overactivity and a reduction in the apparent need for sleep may be interpreted by family and carers as a behavioural side-effect of the new drug. In fact it is merely a consequence of the person's personality being unleashed from the effects of the epilepsy. In people with learning disabilities who have been impaired by epilepsy for years, this sudden unleashing may present an adjustment problem. It is not a side-effect of the anti-epileptic drug as such, it is really a confirmation of a therapeutic effect.

In order to assess the relationship between drug treatment, epilepsy and behaviour change, it is therefore essential to consider the following in addition to the usual enquiries about dose, relationship of event in time to use of the drug, and so on:

(a) the type of epilepsy;
(b) the previous history of behavioural change (if any);
(c) the relationship of symptoms to changes in seizure frequency and spectrum of seizure types; and
(d) other possible aetiologies for the change.

Other aspects of treatment

Psychological treatments

Psychological treatments may be used to manage intercurrent psychological problems in epilepsy. There is, however, a burgeoning literature on the use of behavioural treatments directly to control seizures. Standard behaviour therapy techniques can be employed (Dahl, 1992; Goldstein, 1997).

Environmental aspects

Seizure frequency can be affected by the emotional climate of a person's carers. Any programme of treatment must include education of carers and family about the nature of epilepsy and its treatment. Contact with one of the voluntary organisations in epilepsy may be helpful (such as the British Epilepsy Association, which has a freephone helpline 0800 309030). Perhaps the biggest single problem that people with epilepsy face is the danger of overprotection. Although life has dangers, quality of life depends on being able to take a measured assessment of its risks.

Service issues

Liaison with primary care teams

There is some evidence that specialist services for epilepsy are more effective than neurology services. In some parts of the UK joint clinics between learning disability specialists and epileptologists have been established. However, the primary health care team will provide the ongoing management of the patient's epilepsy. Effective liaison between the hospital and primary care services can be achieved in a number of ways. Epilepsy specialist nurses may be able to work with the primary care team to insure that information is available for both professionals and patients. Cooperation cards, which may be locally designed and act as a form of patient-held records, are extremely useful in maintaining continuity of treatment between the primary and secondary services.

Epilepsy specialist nurses

One of the most significant developments in the structure of services in the past few years has been

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the emergence of epilepsy specialist nursing, including nurse practitioners in epilepsy. Specialist nurses may liase between primary and secondary care, and between different parts of the secondary care service. They may also be involved with counselling and group work. Evaluation of the effectiveness of epilepsy nurse specialists has been attempted and is still ongoing (Taylor *et al*, 1994; Appleton & Sweeney, 1995). It does seem likely, however, that this new speciality will continue to expand and develop in the future.

Conclusions

Seizure disorders are common in people with learning disabilities, and present special challenges in diagnosis and treatment. Psychiatrists in learning disabilities have a special part to play in management of these conditions, and should be comfortable about doing so. Proper use of new drug treatments may be particularly important, and physicians should not shirk from ensuring that their patients get proper access to relevant investigations and treatment. The drug treatment of epilepsy may have behavioural consequences other than drug side-effects, and a diagnosis of the cause of behaviour change should be sought. The drug treatment of epilepsy is part of an overall package, which should include attention to social and environmental factors.

References

Anzola, G. P. (1993) Predictivity of plasma prolactin levels in differentiating epilepsy from pseudoseizures: a prospective study. *Epilepsia*, **34**, 1044–1048.

Appleton, R. E. & Sweeney, A. (1995) The management of epilepsy in children: the role of the clinical nurse specialist. *Seizure*, **4**, 287–291.

Brown, S. W. (1993) Management issues in treating adults with lamotrigine – II. In *Lamotrigine – A New Advance in the Treatment of Epilepsy: Royal Society of Medicine International Congress and Symposium Series No. 204* (ed. E. H. Reynolds), pp. 95–100. London: Royal Society of Medicine.

—, Betts, T., Chadwick, D., *et al* (1993) An epilepsy needs document. *Seizure*, **2**, 91–103.

Cassidy, G. & Corbett, J. (1997) Learning disorders. In *Epilepsy: A Comprehensive Textbook* (ed. J. Engel Jnr & T. Pedley), pp. 2053–2063. Philadelphia, PA: Lippincott-Raven.

Collins, W. C., Lanigan, O. & Callaghan, N. (1983) Plasma prolactin concentrations following epileptic and pseudoseizures. *Journal of Neurology, Neurosurgery and Psychiatry*, **46**, 505–508.

Dahl, J. (1992) *Epilepsy: A Behaviour Medicine Approach to the Assessment and Treatment in Children. A Handbook for Professionals Working with Epilepsy*. Gottingen: Hogrefe and Huber.

Deb, S. & Hunter, D. (1991*a*) Psychopathology of people with mental handicap and epilepsy: I. Maladaptive behaviour. *British Journal of Psychiatry*, **159**, 822–826.

— & — (1991*b*) Psychopathology of people with mental handicap and epilepsy: II. Psychiatric illness. *British Journal of Psychiatry*, **159**, 826–830.

Espie, C. A., Pashley, A. S., Bonham, K. G., *et al* (1989) The mentally handicapped person with epilepsy: a comparative study investigating psychosocial functioning. *Journal of Mental Deficiency Research*, **33**, 123–135.

Fenwick, P. (1995) Psychiatric disorder and epilepsy. In *Epilepsy* (2nd edn)(eds A. Hopkins, S. Shorvon & G. Cascino), pp. 453–502. London: Chapman & Hall.

Goldstein, L. H. (1997) Psychological control of seizures. In *The Clinical Psychologist's Handbook of Epilepsy: Assessment and Management* (eds C. Cull & L. H. Goldstein), pp. 113–129. London: Routledge.

Hopkins, A. & Shorvon, S. (1995) Definition & epidemiology of epilepsy. In *Epilepsy* (2nd edn) (eds A. Hopkins, S. Shorvon & G. Cascino), pp. 1–24. London: Chapman & Hall.

International League Against Epilepsy Commission on Classification and Terminology (1989) Proposal for revised classification of epilepsies and epileptic syndromes. *Epilepsia*, **30**, 389–399.

International League Against Epilepsy Commission on Epidemiology and Prognosis (1993) Guidelines for epidemiological studies on epilepsy. *Epilepsia*, **34**, 592–596.

Jacoby, A., Baker, G. A., Steen, N., *et al* (1996) The clinical course of epilepsy and its psychosocial correlates: findings from a UK community study. *Epilepsia*, **37**, 148–161.

Jenkins, L. K. & Brown, S. W. (1992) Some issues in the assessment of epilepsy occurring in the context of learning disability in adults. *Seizure*, **1**, 49–55.

Kanemoto, K., Kawasaki, J. & Kawai, I. (1996) Postictal psychosis: a comparison with acute inter-ictal and chronic psychoses. *Epilepsia*, **37**, 551–556.

Logsdail, S. J. & Toone, B. K. (1988) Post-ictal psychoses. *British Journal of Psychiatry*, **152**, 246–252.

Mattson, R. H. (1996). The role of the old and the new antiepileptic drugs in special populations: mental and multiple handicaps. *Epilepsia*, **37** (suppl. 6), S45–S53.

Moore, P. M., Baker, G. A., McDade, G., *et al* (1994) Epilepsy, pseudoseizures and perceived family characteristics: a controlled study. *Epilepsy Research*, **18**, 75–83.

Nashef, L. & Brown, S. (1996) Epilepsy and sudden death. *Lancet*, **348**, 1324–1325.

Rutter, M., Graham, P. & Yule, W. (1970) *A Neuropsychiatric Study in Childhood*. London: Heinemann Medical.

Scheepers, B., Budd, S., Curry, S., *et al* (1994) Non-epileptic attack disorder: a clinical audit. *Seizure*, **3**, 129–134.

Scheffer, I. E., Bhatia, K. P., Lopes-Cendes, I., *et al* (1995) Autosomal dominant nocturnal frontal lobe epilepsy. A distinctive clinical disorder. *Brain*, **118**, 61–73.

Smith, M. C. (1997) Landau-Kleffner syndrome and continuous spikes and waves during slow sleep. In *Epilepsy: A Comprehensive Textbook* (eds J. Engel Jnr & T. Pedley), pp. 2367–2377. Philadelphia, PA: Lippincott-Raven.

Staufenberg, E. F. & Brown, S. W. (1994) Some issues in non-convulsive status epilepticus in children and adolescents with learning difficulties. *Seizure*, **3**, 95–105.

Steffenburg, S., Gillberg, C. & Steffenburg, U. (1996) Psychiatric disorders in children and adolescents with mental retardation and active epilepsy. *Archives of Neurology*, **53**, 904–912.

Taylor, M. P., Readman, S., Hague, B., *et al* (1994) A district epilepsy service with community-based specialist liason nurses and guidelines for shared care. *Seizure*, **3**, 121–127.

Trimble, M. R. (1991) *The Psychoses of Epilepsy*. New York: Raven Press.

Trimble, M. R. & Schmitz, B. (1997) The psychoses of epilepsy/schizophrenia. In *Epilepsy: A Comprehensive Textbook* (eds J. Engel Jnr & T. Pedley), pp. 2071–2081. Philadelphia, PA: Lippincott-Raven.

Multiple choice questions

1. In North America and Europe the lifetime risk of developing epilepsy is:
   a one in 200
   b one in 300
   c one in 30
   d one in 10.

2. Which of the following statements is true:
   a epilepsy is a lifelong condition
   b the ability to have seizures is present in all normal living brains
   c epilepsy carries an excess risk of suicide
   d an EEG must be carried out to confirm that the diagnosis is epilepsy.

3. Complex partial seizures:
   a are similar to simple partial seizures but of longer duration
   b include Jacksonian motor epilepsy
   c always involve impairment of consciousness
   d may resemble absence seizures in their presentation.

4. Lennox–Gastaut syndrome:
   a usually starts in the teenage years
   b is commonly associated with learning disability
   c responds best to carbamazepine
   d is commonly associated with non-convulsive status epilepticus.

5. Non-epileptic seizues:
   a are usually a form of malingering
   b often arise in sleep
   c often may represent a form of post-traumatic stress disorder
   d are characterised by a post-ictal rise in serum prolactin.

MCQ answers

1	2	3	4	5
a F	a F	a F	a F	a F
b F	b T	b F	b T	b F
c T	c T	c T	c F	c T
d F	d F	d T	d T	d F

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