Chapter 41

Epilepsy and learning disability

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Definition

Learning disability is defined as a composite of:
1. Deficiency in learning (Intelligence Quotient (IQ) less than 70)
2. Difficulties with daily living skills
3. An onset within the developmental period (less than 18 years of age).

Epidemiology

Epidemiological issues in 'special groups' are dependent on both the source and age of the population. Cohort effects, due to year of birth, are important in defining prevalence in both learning disability[1] and epilepsy[2]. Table 1[3–7] shows epidemiological surveys of the prevalence of epilepsy in people with mental and physical handicap. A survey in an institution for people with learning disability gave a prevalence of epilepsy of 32%[5], while a large community-based questionnaire survey of health needs in people with a learning disability gives a prevalence of 22.1%, making epilepsy second only to psychological illness as a comorbidity[7]. This can be compared with an estimated prevalence of epilepsy in the general population of between 0.4 and 1%[8].

Table 1. Epidemiological surveys of the prevalence of epilepsy in people with mental and physical handicap[3-7].

Study	Sample	Prevalence
Corbett et al (1975)[3]	Children under age 14 Community SMR	 20%
Richardson et al (1981)[4]	Children up to 22 yrs Community MMR SMR	 24% 44%
Mariani et al (1993)[5]	Institution	32%
Steffenburg et al (1995)[6]	Children 6–13 year old Community MMR SMR	 14% 24%
Welsh Office (1995)[7]	Adults Community-based All MR	 22.1%

MMR: mild mental retardation, IQ 50–70; SMR: severe mental retardation, IQ <50

Seizure type and seizure syndrome

A community study of children with learning disability[9] reflected on the difficulties of defining seizure type. This was because only 10% of the population with severe physical and mental handicap underwent electrophysiological tests in this study. The authors showed an increase in generalised tonic-clonic and myoclonic seizures and a decrease in partial seizures with increasing handicap and concluded that this increase in generalised seizure disorder was an artefact of the lack of investigation in this population, though other explanations such as genetic causes may be valid.

In an institutionalised population Mariani and colleagues[5] showed 32.5% of subjects to have partial epilepsy and 62.5% to have generalised epilepsy, with 5% unclassified. Interestingly, in the population with generalised epilepsy, 31.4% had EEG changes typical of idiopathic epilepsy. Unfortunately further data on seizure type or syndromal diagnosis in these patients was not given. It seems from these two sources that generalised abnormalities, and hence appropriate treatment options, should not be unexpected in people with learning disability.

Assessment

Aetiological factors

Learning disability is caused by a range of pathological processes, as of course is epilepsy itself. The underlying cause of the learning disability has an impact on seizure type and outcome.

Epilepsy phenotypes

The seizure disorder associated with some conditions, for example tuberous sclerosis[10], has been well defined. In the case of tuberous sclerosis the value of a good epidemiological survey was shown with a lower than expected prevalence of learning disability in the condition than previously recognised. The nature of epilepsy in Down syndrome has been characterised[11]. A seizure disorder is often associated with Alzheimer's disease, particularly if onset occurs over 30 years of age. This obviously has a significant impact on the outcome of new onset epilepsy in this age group.

For some other conditions associated with disability, such as the fragile X syndrome, epilepsy conditions specific to the syndromes have been suggested. In the case of fragile X there are reports that a specific EEG abnormality similar to benign childhood epilepsy with centro-temporal spikes is present[12] although controversy remains over the validity of this finding – possibly due to sampling and other methodological issues[13]. Table 2 summarises these epilepsy phenotypes[10–12,14]. Rett syndrome poses a specific challenge. The condition is associated with high levels of epilepsy, possibly as a result of the frequently severe level of intellectual disability. However the condition can also offer diagnostic challenges with the frequent hyperventilation and other autonomic disturbances being misdiagnosed as partial or other seizure types.

Other impairments

The association between the likelihood of having epilepsy if an individual has an additional impairment is strong. Hauser and colleagues[15] showed an increase in the risk of epilepsy from 11% to 48% when a child with learning disability also had cerebral palsy – an association confirmed by others[16]. Steffenburg and colleagues[6] showed a prevalence of cerebral palsy of 14% and 59% respectively in the mild and severe groups of patients with learning disability and epilepsy. In the population with learning disability who had epilepsy the risk of additional impairment was 3% in the population with mild disability and 37% in those with severe disability.

Table 2. Suggested epilepsy phenotypes in genetic conditions causing mental handicap[10-12,14].

Condition	Nature of epilepsy, provisional	Study
Angelman syndrome	Seizure onset in early childhood, evolution of seizure type from high-voltage slow bursts in infancy to diffuse spike and wave in middle childhood. Atypical absences and absence status	Matsumoto et al (1992)[14]
Tuberous sclerosis	62% risk of developing seizures	Webb et al (1991)[10]
Fragile X syndrome	Debate over specific EEG changes similar to benign childhood epilepsy with centro-temporal spikes	Musumeci et al (1991)[12]
Down syndrome	Seizure prevalence of 1–13%. Two peak incidences in first year of life and later life, the latter being associated with the presence of Alzheimer's disease	Stafstrom (1993)[11]

In addition to complex physical and sensory impairments this population has a high prevalence of other co-morbidities. Communication difficulties are inevitable and will lead, as we shall discuss, to difficulties in the diagnostic and treatment process. It is however the high prevalence of behaviour disorder, with an estimated community prevalence for psychiatric and emotional disturbance of 32.2% in people with learning disability[7], that can affect both assessment and treatment. This leads to two main confounders. First, confusion of behaviours not associated with epilepsy with those that are epilepsy related and, second, the effect of prescribing antipsychotic medication, due to their known epileptogenic potential[17]. Many studies have looked at the prevalence of antipsychotic medication in populations of people with learning disability[18]. Prevalence figures range from 40.2% in hospitals, through 19.3% in the community, to 10.1% in family homes.

Diagnosis

Communication skills – management by proxy

As mentioned previously, the complexity of aetiology and the presence of communication difficulties alters our approach and may diminish reliability. The ability to communicate and place at ease the individual with learning disability is a key skill for any epileptologist. It is known, for example, that young people with profound learning disability can discriminate between familiar people and those who are strangers, and are able to form personal relationships. When inexperienced strangers try and communicate with this group of people they have significantly less interactive and communicative involvement[19]. Unfortunately many doctors have little training in this area.

In people with learning disability, a witness report from a carer or family member is common, a report from the individual is less so. Thus our history and management will commonly progress through another – 'management by proxy'. The degree of this will increase as the individual's communicative skills decrease.

Good quality communication skills can be achieved through education. Analysis of communication suggests that addressing the following skills would be appropriate:

1. Non-verbal; gaze, appropriate touch, use of gesture
2. Vocal; appropriate tone, intelligibility
3. Verbal; greeting, using individual's name, balance of communication with carer

4. Response; recognising the individual's responses and following leads, respecting information from care giver
5. Empathy; showing appropriate respect and empathy[20].

Specific issues in differential diagnosis: seizures or behaviour disturbance?
In the majority of cases seizure disorder presents itself as paroxysmal episodes of abnormal behaviour. In many cases, a generalised tonic-clonic convulsion for example, the nature of these behaviours is well defined and does not mimic many other conditions. Other seizure disorder, however, is less well defined or is dependent on the verbal description of the individual and witnesses for a diagnosis. An example of the former is the pattern of behaviour seen in complex partial seizures, particularly when there are associated ictal or post-ictal automatisms. Differentiating these in the general population from psychiatric disturbance or, in some cases, from non-epileptic attack disorder is complex. Differentiating these in people with learning disability is further complicated by communication issues and the high prevalence of behaviour and motor disorders in this population.

Repetitive episodes of manneristic or stereotyped behaviour would be most unusual in many people without handicaps and the diagnosis of epilepsy would be highly likely. However in a young man with autistic tendencies, for example, such behaviours may be reflections of the cognitive disturbance of the autism and not in fact epilepsy. Clinicians need a structured approach to this differentiation. Table 3 highlights guidelines to this differential diagnosis, though in many cases behavioural analysis will be required to sufficiently differentiate the behaviour.

Treatment

Unfortunately people with learning disability do not fit well into established evaluation processes. This can be seen by a continued trend to open trials and retrospective case note evaluations with a paucity of randomised, controlled trials, as we will discuss later.

Table 3. Differentiating seizure and behaviour disorder.

Seizure	**Behaviour disturbance**
Identical behaviour on each occasion	Variation in behaviour with circumstances
No precipitant	Commonly precipitant such as demands, need to avoid situation
Unresponsive to communication, calming	Responsive to calming, support, removal from stressor
Investigations: Analysis of behaviour: no relationship to behaviour and environment Video: Shows typical seizure features EEG: positive inter-ictal EEG	*Investigations*: Analysis of behaviour: relationship found. Video: Atypical picture seen EEG: negative inter-ictal EEG of some use

In clinical practice with people with learning disability we are left with something of a clinical effectiveness dilemma. To practice purely by gold-standard approaches leaves us with precious few interventions, and almost zero comparative studies. We therefore apply knowledge on interventions gained in the general population to this special population, but the validity of this approach in this population remains unproven, in particular for assessment of side effects. The latter course of action is, of course, a clinical necessity.

Clinical effectiveness data in people with learning disability

Studies looking at this population have been divided into assessing practice, usually antiepileptic drug (AED) reductions through cohort or intervention studies, and pharmacological interventions to control seizures.

Cohort studies and drug reduction

Cohort studies looking at practice over several years have been performed in institutional[21,22] and clinic populations[23,24]. Pellock and Hunt[22] reviewed ten years of treatment in an American institution using an open methodology and showed a trend towards reduction in polytherapy (19%), with a relative increase in monotherapy and a large decrease in patients receiving three anticonvulsants (a decrease of 47.6%), and a decrease in the use of barbiturate anticonvulsants. Poindexter and colleagues[21] showed a similar trend towards medication rationalisation and in particular reduction of barbiturate anticonvulsants. Singh and Towle[23] followed 100 patients with learning disability over a mean duration of 7.5 years in an outpatient referral setting. This survey is an interesting reflection on clinical practice with 60% of patients maintained on one, 38% on two, and 2% on three AEDs. Tobias and colleagues[24] audited the practice of a large British outpatient epilepsy service through 1000 consecutive referrals. Again, essentially through a cohort study, it enabled comparison between people with and without handicap and shows that there was a trend toward withdrawal of barbiturate anticonvulsants in the general population over this period.

Several intervention studies have assessed drug reduction or 'rationalisation'. Fischbacher[25] showed in an uncontrolled or randomised study that reduction of at least one AED was feasible for many patients and could have an associated behavioural improvement. Beghi and colleagues[26], using a similar uncontrolled non-randomised approach, were able to show a reduction in AEDs from 1.84 to 1.05 per patient over a mean of 12.5 months. A further non-controlled, open, non-randomised study from the UK[27] showed that out of 172 patients remaining over three years (from a population of 215 patients) the mean number of AEDs reduced from 1.41 to 1.05 per patient. This was associated with an increase in dosage of remaining drugs and a less than clear effect on seizure frequency, with a reduction in 48% of patients, an increase in 33% and no change in 19%. Unfortunately for the practising clinician, while there appears to be a groundswell of support for 'rationalisation', aspects of the methodology used in all of the above studies, crucially lack of control and randomisation, leave the issue unproved.

Some guidance for the clinician intending to discontinue medication when a patient has been seizure free can be gained from the work of Alvarez[28]. In a non-randomised, controlled, but well described study the author showed, with an impressive eight-year follow-up period, that following a seizure-free period of at least two years an attempt at reduction could be made. In this population of 50 patients seizures recurred in 26 (52%); 11 of these occurred during discontinuation and 30% after discontinuation. A total of 80% of recurrences occurred less than three years after the start of discontinuation. Predictors of successful discontinuation are (1) few documented seizures in a lifetime, (2) no gross neurological abnormalities, (3) low drug levels at initial discontinuation, and (4) persistently normal EEGs before and after discontinuation.

Pharmacological interventions

The majority of data on pharmacological studies, with some notable exceptions to be discussed later, concern add-on, open, non-randomised design, usually with the novel AEDs. Such studies are reasonably numerous but, of course, are open to methodological criticism and hence interpretation is difficult.

Trials using open non-controlled methodology in populations with learning disability and refractory epilepsy have shown a 50% reduction in seizures in 33% of patients at three-month follow up on vigabatrin[29], with a reduction in this response by one-third at five-year follow-up[30].

A similar methodology using lamotrigine in a childhood population[31] showed a 50% improvement in seizure control in 74% of children, with an associated improvement in quality of life using clinical judgement.

In addition to these studies, which have tended to investigate cohorts of individuals with learning disability, a further fruitful area of pharmacological research has been in epilepsy syndromes strongly associated with learning disability – West syndrome, infantile spasms, and the Lennox-Gastaut syndrome. The former, being a developmental age-defined syndrome, is somewhat less useful in the population we are studying, however. Chiron and colleagues[32] have shown in both open and a limited placebo-controlled run-in an impressive efficacy for vigabatrin in this population, with 43% of children showing complete cessation of seizures and 46 out of 70 children showing a greater than 50% reduction in seizures. In a recent report, in abstract form, of a double-blind, placebo-controlled study of vigabatrin in infantile spasms Appleton and Thornton[33] showed a complete cessation of seizures in 45% of the active versus 15% of the control group.

The clinical effectiveness data in Lennox-Gastaut syndrome is of particular interest to clinicians dealing with both children and adults with learning disability. Two good quality randomised controlled trials have been performed.

Lamotrigine has been subject to the most rigorous quality of life evaluation in the Lennox-Gastaut population. The compound has been investigated through a randomised, placebo-controlled, add-on design[34]. Importantly, however, this study used a specifically designed quality of life scale and parental global health evaluation in addition to the usual seizure frequency measures. In terms of seizure efficacy the study was successful with a significant reduction in atonic seizures and in total seizures. The impact on quality of life measures was interesting. Parent/carer assessment showed an improvement in global health. Outcome on the ELDQOL showed significant improvement in mood and reduced seizure severity, but no difference in side effect profile was seen when compared with placebo.

Topiramate. This study recruited 98 patients aged 2–42 years. Primary successful outcome points were deemed to be either a combination of a significant reduction in atonic (drop) attacks and parental global evaluation of seizure severity or a percent reduction of all seizure types. It can be seen that some attempt was made to evaluate the impact on quality of life through these parental evaluations.

The methodology applied was a randomised, placebo-controlled, add-on design. The population had quite severe seizures with all having at least 60 seizures per month.

Results showed a statistically significant median reduction in drop attacks (placebo increased by 5%, topiramate decreased by 15%; $P = 0.04$) and in parent evaluation of seizure severity (placebo 28% improvement, topiramate 52%). There was no statistically significant decrease

in overall median seizure frequency[35]. Parental global seizure severity was the only chosen measure of quality of life in this study.

A further study[35] used a RCT approach to add-on therapy in adults with learning disability and epilepsy. This study showed a reduced seizure frequency of >30% in the topiramate group as compared with 1% in placebo ($P = 0.052$).

Levetiracetam has not been trialled in a RCT design within this population. In an open study[36] 64 patients were given add-on levetiracetam after a three-month baseline. In this study 24 patients (38%) became seizure free and there were a further 18 responders (28%).

Pregabalin and zonisamide are relatively new to the market and it is expected that similar case review studies will be seen soon.

Rufinamide has been studied in an RCT[37] in which 138 randomised patients received rufinamide or placebo. Significant improvements were seen in total seizure frequency, 'drop-attacks' and a higher 50% responder rate. Common adverse events included somnolence and vomiting.

Further details on both pharmacological and non-pharmacological studies can be seen in two Cochrane reviews (please see Further reading).

Treatment choice

The decision of treatment choice for people with learning disability is broadly split into two components. Firstly, choice should be based on seizure type, seizure syndrome, individual patient characteristics and patient and carer choice. Patients and carers will have specific concerns over drugs that may have cognitive or behavioural side effects. The clinician should clearly describe these potential effects when informing patients. This can be a major concern in those with co-existing behavioural problems, which can be at least 40% of the adult population

Secondly, the clinician should assess remaining treatment options. People with learning disability will often be on multiple therapies and will have tried several AEDs. It is important to place a patient on a treatment pathway to assess what available untried epilepsy options are available, whether previous options can be retried, and whether the current treatments can be removed or dosage changed. A simple checklist for a clinician would be:

1. Current therapy. Can any of the AEDs be increased without unwanted side effects? This is particularly useful if the AED has shown some evidence of efficacy. If on polytherapy, can a drug be removed?

2. If none of the above, has the patient had all the available AEDs, including 'new' AEDS such as: lamotrigine, levetiracetam, pregabalin and topiramate?

3. If a patient has focal seizures, has assessment for resective surgery been considered?

4. If patient has tried all AEDs and is not candidate for resective surgery, has assessment for vagal nerve stimulation been considered?

Making your treatment work

Applying treatment should be relatively easy in that many people with learning disability will have carers who can aid in giving the treatment. The clinician will need to ensure that carers are capable of giving medication and should also identify whether the patient has any swallowing problems and can take the formulation prescribed. As a general rule caution in

dose escalation is recommended; *start low go slow* is a reasonable policy and usually very acceptable to carers. In fact, it is not uncommon to prescribe drugs in the lowest available doses, building up slowly to recommended treatment doses.

Outcome assessment is more complicated. Due to the refractory nature of epilepsy and concerns over side effects, treatment outcome frequently focuses on assessing the relative value of any seizure change and judging any potential negative impact of AEDs. The ideal is to establish outcome goals *prior to initiating treatment*, though unfortunately we often have to assess outcomes retrospectively. Thus decisions should be made pre-treatment to appropriate seizure outcomes. Seizure freedom remains the goal; however significant seizure reduction, reduction in specific harmful seizures (such as atonic seizures) or changes in cognition may all be goals of treatment.

Seizure counting is important. However, specific help will be needed to count each type of seizure accurately. It may be very hard to assess alteration in absences. Side effects can be very difficult to judge. In particular, altered behaviour is likely to be related to behavioural problems already present pre-treatment. Behaviour change can also occur when seizures are reduced (so-called forced normalisation), and this is best approached by managing any change in behaviour through local support services.

To avoid leaving the patient on an increasing number of AEDs it is also good practice to come to a decision on whether the treatment change has been successful, and if it has not then the new treatment should be removed.

Special issues: assessing the interaction of behaviour and epilepsy

As we have already discussed the interaction between behaviour and epilepsy is important, not solely for differential diagnosis but also because side effects of treatment may often have behavioural presentations.

Figure 1 sets out guidelines for the clinician to assess the relative likelihood that a behaviour is linked to epilepsy or its treatment. The key element of this assessment is the ability to describe the meaning of the behaviour, the so-called 'functional analysis of behaviour'. This may in fact need to be done with such a degree of sophistication that referral to, and working with, community nurses or psychological services will be necessary. With this a clinician should be able to assess whether a particular behaviour is in fact caused by seizures, caused by medication or independent of both seizures and medication.

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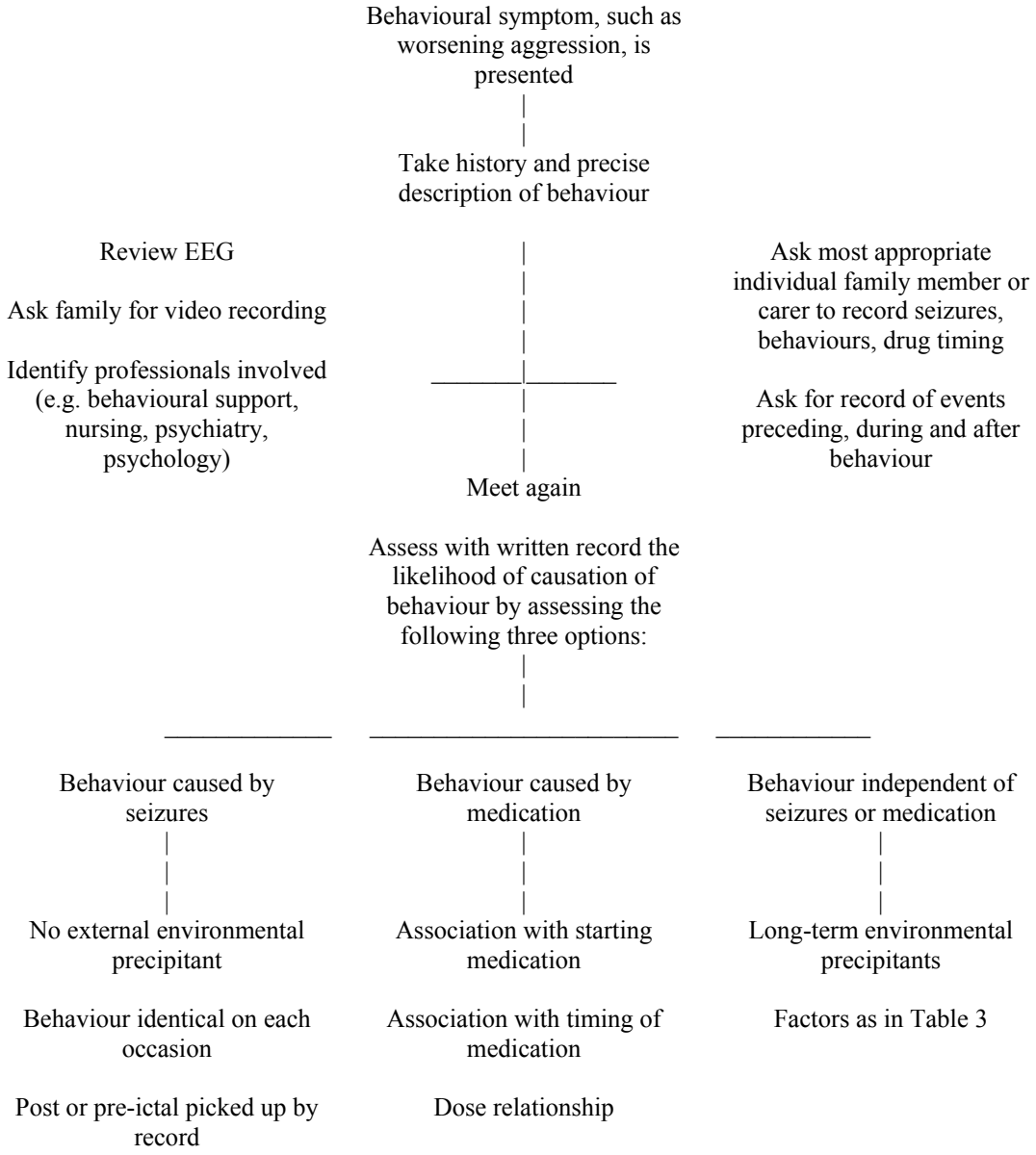

Figure 1. Assessing behavioural symptoms in epilepsy in people with learning disability.

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