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Epilepsy in adults with learning disabilities

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Epilepsy is a common problem of adults with learning disabilities. Despite the high prevalence there have been few studies of the epilepsy suffered by adults with learning disabilities. The findings of a Leicestershire study are that multiple seizure types are a common presentation, and that for many (75%) the seizures remain refractory to treatment. Those who suffer tonic–clonic seizures are most likely to achieve remission, whereas for those with simple or complex partial seizures the prognosis is poor. For a significant proportion, status epilepticus, emergency admission to hospital and injuries occur on a regular basis.

Key words: epilepsy; learning disabilities; seizures; epidemiology; status.

INTRODUCTION

Most of the key epidemiological studies of epilepsy and learning disabilities have involved children, rather than adults[1–3]. Corbett *et al.*[1] found that of children with an IQ lower than 50, one-third had a history of seizures and 19% had had at least one seizure during the previous year. A study of Aberdeen[2] showed that by the age of 22, 15% of children with learning disabilities had epilepsy and an additional 7% had experienced at least one seizure. Children with a history of post-natal injury suffered a much higher risk of epilepsy. The study of a whole population of people with learning disabilities in a northern Swedish county found 20.2% to suffer from epilepsy[3]. Despite this high prevalence of epilepsy there are remarkably few studies of adults with learning disabilities.

Goulden *et al.*[2] reported that by the age of 22 years 39% had achieved 5 years free of seizures and this performance varied according to the nature and cause of the learning disabilities. For those with no associated disability a 5-year seizure-free remission was achieved in 56% of cases. Remission was also reported in 47% of cases with associated cerebral palsy but only 11% of those with post-natal injury. They concluded that epilepsy in the population will frequently enter remission in later life.

The natural history of epileptic seizures shows that the greater incidence of new cases appears between the ages of 1 and 4 years[4]. Similar results were shown in a study of a learning disabled population living in institutional care in Norway[5]. A primary concern of that study was that many people in prolonged remission remained on antiepileptic drugs, which had central nervous system side effects.

The presence of both epilepsy and learning disabilities are indicators of both early mortality and psychiatric disorder. A population-based 35-year follow-up study of 245 subjects with childhood-onset epilepsy showed that 49% of the subjects had learning disabilities. In addition, 86% of those that had died had suffered learning disabilities[6]. Steffenburg *et al.*[7] studied a cohort of 98 children with active epilepsy and concluded that a majority of those with both epilepsy and learning disabilities suffered from a psychiatric disorder.

Many of these studies might lead one to believe that the sole aim in relation to epilepsy in adults with learning disabilities is to continue the treatments successfully started by paediatricians and to withdraw treatments when appropriate. For many adults with learning disabilities, though, the occurrence of epileptic seizures still remains a major problem. Forsgren *et al.*[3] showed the prevalence of epilepsy in learning disabilities remaining constant during much of the period from 20–60 years, while one study showed as many as 29% suffering a first seizure after the age of 20[5]. In addition, for many the epilepsy is likely to be a confounder in a complex presentation of behaviour disorder, mental illness and epilepsy.

The lack of knowledge about the epilepsy of adults

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with learning disabilities led to the current study. Concern about the lack of efficacy of new drug treatments[8] and the apparent refractory nature of the epilepsy suffered by so many people underlined the lack of knowledge. This paper reports the findings of a study to provide an overview of the epilepsy suffered by adults with learning disabilities living within a defined geographical area.

MATERIALS AND METHODS

The County of Leicestershire, UK, encompasses the City of Leicester and a number of county towns (total population approx. 900 000 people). Approximately 2600 adults with learning disabilities are known to the learning disabilities register and this is estimated to be in excess of 90% of adults with learning disabilities who live in Leicestershire and receive services either from health or social services.

The register contained information about the likelihood of a person suffering epilepsy from six sources. These sources included a hospital census undertaken in 1988, a Leicestershire-wide programme of medical examinations, community nurse records, two drug surveys[9] and other core data from other databases. The most comprehensive source of information was the 5-year rolling programme of carer interviews undertaken by the trained interviewers employed by the learning disabilities register. This contained information on all adults with learning disabilities over 24 years of age known to the learning disabilities register and a proportion of those aged 18–24. These various sources provided a total of 796 people who were thought to possibly suffer from epilepsy.

For the purpose of this study a questionnaire was devised. It comprised five main areas of enquiry: does the person suffer from epilepsy, the nature of the epilepsy, the treatment of the epilepsy, the current access to services for epilepsy, and the frequency of injuries sustained and admission to hospital. The questionnaire was piloted on one ward with a large number of patients who suffered from epilepsy, and then subsequently revised.

For the purposes of the study a person who suffers from epilepsy was defined as a person who suffers from epileptic seizures (fits) or has done so in the past. The person can either suffer from epileptic seizures or/and receive antiepileptic drugs to control those seizures. People who were prescribed antiepileptic drugs for other reasons, such as the control of maladaptive behaviour, were not included. Carers were asked to confirm whether or not the person with learning disabilities suffered from seizures. If they were not sure further enquiry took place firstly by a telephone call to the carer, and secondly by scrutiny of the medical notes.

Carers who confirmed that the person suffered from epilepsy were asked to complete questions about the nature and frequency of the seizures. Carers were provided with a series of descriptions of seizures adapted from descriptions provided by the British Epilepsy Association[10] and asked to indicate which types the person suffered from.

The questionnaire was distributed in two phases, the first to hospital inpatients and the second to people who lived in the community. One month after the distribution of the 798 questionnaires a follow-up questionnaire was sent for those that had not returned the original. In addition, a telephone service was provided to assist those that were either having difficulties understanding the questionnaire or required a person to speak another language. The information from the questionnaire was entered on to a computer database and analysed using SPSS.

RESULTS

Of 798 questionnaires, 689 were completed (86%). Of the 689 completed questionnaires, 532 confirmed that the person suffered from epilepsy and 137 stated that the person did not. For the remaining 20, diagnostic confusion remained. All subsequent results are for the 532 people suffering from epilepsy. At the time of the study approximately 2600 adults with learning disabilities were known to the learning disabilities register. Extrapolation of the data of this study would give a prevalence rate for epilepsy of 25% of adults with learning disabilities. The proportion of those with epilepsy remained at approximately 30% between the ages of 20 and 50 but declined steadily after that age. Only 11% of persons over the age of 70 years suffered epilepsy.

Table 1: The frequency of seizure types.

Seizure type	Number of persons suffering seizure type (%)[a]
Tonic–clonic	291 (60%)
Absences	187 (37%)
Myoclonic jerks	99 (21%)
Tonic	76 (14%)
Drop attacks	36 (7%)
Simple partial seizure	67 (14%)
Complex partial seizure	28 (6%)
Complex partial seizure with secondary generalization	37 (8%)
Other description	35 (7%)

[a] Total is in excess of number of respondents due to many people suffering from more than one seizure type.

Demographic details

Of the 532 adults who suffered from epilepsy 296 (56%) were male and 236 were female (44%). Their age distribution was from 18 to 86 years, with 139 (26%) aged between 18 and 29 years, 175 (33%) aged

between 30 and 39 years, 120 (22%) between 40 and 49 years, 58 (11%) between 50 and 59 years while the remaining 40 (8%) were over 60 years of age (mean age = 38.6 years ±12.97).

Information about the degree of learning disabilities was available for 482 of the 532 identified as suffering from epilepsy. Two hundred and thirty (48%) suffered profound learning disabilities, 121 (25%) severe learning disabilities, 99 (22%) moderate learning disabilities and 32 (7%) mild learning disabilities. This distribution was different from that of the whole population known to the register.

The 532 people were living in a wide variety of settings. One hundred and seventeen lived in wards, hostels and group homes managed by the National Health Service (NHS), 176 lived in residential homes of other agencies (including private) and 239 lived at home with parents or relatives.

Of those who suffered epilepsy, for 300 (56%) the questionnaire was completed by a carer, whilst for 214 (40%) it was completed by a relative. For the remaining 13 (2%) the person completing the questionnaire was neither a relative nor a carer (e.g. social worker, friends, appointee, etc.).

For those 117 living in NHS accommodation the person completing the questionnaire had known the person with epilepsy and learning disabilities for an average of 4.1 years (±3.6). For those in other residential homes it was an average of 5.2 years (±5.9) and for those living at home with relatives the person was known for an average of 27.7 years (±13.1).

The sample was predominantly Caucasian (90%) with the remaining 10% being mostly of Asian origin. This is representative of the whole population of people with learning disabilities who live in Leicestershire (Asian, 10.91%).

The nature of the epilepsy

Four hundred and five responders provided information about the age of onset of the seizures. For 65% of those the seizures began before the age of 5, for 30% the onset was between the ages of 5 and 20, whilst for only 34 had the epilepsy begun after the age of 20 years. Of the 34 with adult-onset epilepsy 24 (73%) were male.

Carers and relatives were provided with lay descriptions of eight seizure types, which correspond to those advocated by the British Epilepsy Association, and asked to indicate which corresponded to the seizures suffered by the respective person. Of the seizure types suffered the most common was tonic–clonic. Information about the prevalence of each seizure type is provided in Table 1.

Table 2: The number of seizure types suffered.

Number of seizure types	Number of people (%)
1	247 (46.4)
2	141 (26.7)
3	51 (9.6)
4	24 (4.5)
5 or more	18 (3.3)
Not informed	51 (9.6)
Total	532 (100)

Table 3: The number of people with active epilepsy, epilepsy in remission and inactive epilepsy.

The nature of the epilepsy	Number (%)
Active epilepsy (suffered at least one seizure in last 3 years)	386 (74%)
Epilepsy in remission (no seizures for 3–5 years)	45 (9%)
Inactive epilepsy (no seizures for 5 years)	91 (17%)
Total	522 (100%)

Ten responders did not provide data on seizure frequency.

Many responders indicated that the person suffered from more than one seizure type. Of the 481 respondents who completed the information about seizure types, approximately half stated that the person suffered from more than one seizure type (see Table 2). Absence seizures also occurred in 30%, myoclonic jerks in 20%, tonic seizures in 14% and partial seizures in 13% of those who suffered tonic–clonic seizures. Myoclonic jerks also occurred in 30%, tonic seizures in 16% and partial seizures in 21% of those who suffered absence seizures.

For many, the seizures occurred in clusters (48%), although in only 10% of cases did they always occur in clusters.

It was common for consciousness to be lost during a seizure with only 91 reporting that consciousness was not lost. For most of those who lost consciousness (298) the loss was for less than 5 minutes. In only 59 cases was the loss of consciousness for a longer period of time. Similarly, when confusion occurred following a seizure for most (256 of the 336 claiming confusion occurred) it was for less than 15 minutes. In only 18 cases was the confusion claimed to be in excess of 5 hours. When asked if the seizures occurred according to a regular pattern, 14 (26.3%) claimed there to be. A greater proportion of people with a moderate degree of learning disabilities showed a regular pattern for their epilepsy ($\chi^2 = 11.39$; df. = 3; $P < 0.01$).

When asked about the factors which precipitated seizures a large number of factors were implicated. They included physical illness (142), if excited (164), constipation (82), menstruation (65), forgotten or incorrect medication (64), if behaviour disturbed (73), if environment too noisy and a multitude of other precipitants. Constipation was more commonly stated to be a precipitant for females ($\chi^2 = 7.37$; df. = 1;

Table 4: The number of people with inactive epilepsy, epilepsy in remission and active epilepsy for each of the main seizure types.

Seizure type	No. with (%) inactive epilepsy (%)	No. with epilepsy in remission (%)	No. with active epilepsy (%)	Total (%)
Tonic–clonic	40 (14%)	26 (9%)	225 (77%)	291 (100%)
Absence	10 (5%)	15 (8%)	162 (87%)	187 (100%)
Myoclonic jerks	6 (6%)	9 (9%)	84 (85%)	99 (100%)
Tonics	6 (8%)	3 (4%)	67 (89%)	76 (100%)
Drop attacks	0 (0%)	4 (11%)	32 (89%)	36 (100%)
Simple partial	0 (0%)	2 (3%)	65 (97%)	67 (100%)
Complex partial	1 (4%)	1 (4%)	26 (93%)	28 (100%)
Complex partial with secondary generalization	2 (5%)	5 (14%)	30 (81%)	37 (100%)

$P < 0.005$), and for those with profound learning disabilities ($\chi^2 = 19.85$; df. = 3; $P < 0.01$), whilst excitement was more commonly a precipitant for males ($\chi^2 = 4.25$; df. = 1; $P < 0.05$). Seizures were precipitated by behavioural disturbance and excitement more commonly amongst those with a moderate degree of learning disabilities than those with other degrees of learning disabilities ($\chi^2 = 11.35$; df. = 3; $P < 0.01$ and $\chi^2 = 27.55$; df. = 3; $P < 0.001$). Physical illness was more likely to precipitate seizures for the younger person with epilepsy rather than the older ($\chi^2 = 22.31$; df. = 4; $P < 0.001$).

Carers were asked to provide details about the frequency of the various seizure types and these data enabled each person to be categorized according to whether they were suffering active epilepsy (at least one seizure in the previous 3 years), epilepsy in remission (no seizures for 3–5 years) or the epilepsy was inactive (no seizures for at least 5 years). Only 91 (17%) were categorized as suffering from inactive epilepsy and 45 (9%) from epilepsy in remission. For the remainder (74%) the person continued to suffer from active epilepsy (see Table 3). For those with active epilepsy 200 (51%) reported that the person suffered from more than one seizure per month and of those, 16 (4%) suffered daily seizures.

The performance in achieving remission from seizures with each seizure type is reported in Table 4. Remission was more likely to be achieved by those adults who suffer solely tonic–clonic seizures.

Carers were asked about the frequency of status epilepticus. Seventeen percent of responders reported that the adult with epilepsy and learning disabilities suffered at least one episode of status during 1996, with 5% reporting that it occurred on more than 10 occasions (Table 5).

Carers were asked about the number of times such uncontrolled seizures resulted in emergency hospital admission during the previous 5 years. In 61 cases such an event had occurred and for 23 people it had occurred more than once. When asked whether injuries had occurred as a direct result of the epilepsy during 1996, of the 113 who stated that injuries had occurred 94 (83%) stated that they were minor, while the remainder stated that they were major. In addition, in 14 cases these injuries occurred on a frequent basis.

Table 5: The number of adults with epilepsy and learning disabilities who suffered a status during 1996.

The number of statuses to occur during 1996	The number of adult with epilepsy learning disabilities (%)
None	389 (83%)
1–5	48 (10%)
6–10	12 (2%)
More than 10	22 (5%)
Total	471 (100%)[a]

[a] Sixty-one respondents did not complete this question.

DISCUSSION

This paper reports on one of the few studies of adults with learning disabilities and epilepsy. The high rate of return of the questionnaire and the comprehensive nature of the Leicestershire learning disabilities register enable good estimates of the likely prevalence of epilepsy in the adult learning disability population.

In this Leicestershire study the prevalence rate of epilepsy amongst the population of adults with learning disabilities was 25%. Although this is a higher rate than that reported by others[1–3, 11], the exact figure will inevitably vary according to the criteria for acceptance onto a learning disabilities register and the definition of epilepsy. For example, Corbett *et al.*'s[1] study included children who had suffered a seizure in the previous year and if a similar criteria had been chosen in this Leicester study both results would have been very similar. The learning disabilities register in Leicestershire is similar to many other registers, in that many people with a mild degree of learning disabilities are not notified as they have no requirement for health or social services.

The decline in the proportion of the adult population with learning disabilities suffering epilepsy after the age of 50 could be the result of two factors. The first is an increased risk of early mortality associated with epilepsy and learning disabilities. A second could be that if the epilepsy was associated with childhood or early adulthood the records may have been lost with time. A 35-year follow-up of 245 persons with epilepsy found that 86% of those that died prematurely suffered from learning disabilities[6]. This would support early

mortality being the likely cause, particularly as one of the primary finding of this Leicestershire study is the refractory nature of the epilepsy. Even this apparent decline could be subject to other factors such as the reduced likelihood of people with profound learning disabilities surviving to old age. Forsgren *et al.*[12] found that although there was an increased mortality in people with epilepsy and learning disabilities that the cause of death was other illnesses such as respiratory infections, heart failure and stroke. What can be concluded from the prevalence figures, though, is that the epilepsy often remains a common problem throughout the years of adult life.

A study in Norway[5] found that 29% of people with learning disabilities suffered their first seizure after the age of 20 years and this was associated with the onset of Alzheimer's disease in Downs Syndrome sufferers. This was not the finding of this Leicestershire study, where adult-onset epilepsy only occurred in 7% of cases. The causes of the late-onset epilepsy were varied, and included exposure to psychotropic medication. There was no increase in the incidence of epilepsy in old age as is seen in the general population[13].

Those with epilepsy were significantly more profoundly learning disabled than those without. This finding is again common to most other studies. Interestingly, those with epilepsy are predominantly looked after by relatives rather than in institutional care. This contrasts markedly with those who present with challenging behaviours who are more commonly cared for in institutions. This is similar to a previous study[9].

One of the most interesting findings of this study was that almost half of the adults with epilepsy and learning disabilities suffered more than one seizure type. Other authors[2, 14] have noted that although tonic–clonic seizures are the most common they are often as a result of secondary generalization and associated with partial epilepsy. For carers of people with learning disabilities there may be some difficulty in differentiating between a primary generalized tonic–clonic seizure and one that is secondarily generalized, particularly if they are infrequent or occur at night.

The main finding of this study is the refractory nature of the epilepsy. In almost three-quarters of cases the person continued to suffer from seizures despite drug treatment. This finding is similar to that of Forsgren *et al.*[3] who found that only 32% were seizure-free in the previous year, but rather worse than the study of children with epilepsy and learning disabilities undertaken in Aberdeen[2]. They found that 39% had achieved 5-year seizure-free remission by the age of 22 years. The study of the refractory nature of individual seizure types is in contrast to that achieved by the general population with epilepsy. Juul-Jenson and Foldspang[4] found the seizure-free rates to be 47% for primary generalized tonic–clonic seizures, 52% for absence seizures, 28% for juvenile myoclonic epilepsy and 49% for partial epilepsy with secondary generalization. Adults with learning disabilities fare far worse.

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