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CASE REPORT

A missed diagnosis. A missed opportunity for community integration

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The quality of life of people with learning disability and epilepsy, in the community, can be improved by regular review and appropriate use of anti-convulsants. This paper reports two successful outcomes using a newer anti-convulsant, lamotrigine.

Key words: poorly controlled epilepsy; lamotrigine; learning disability; non-convulsive status epilepticus.

Epilepsy is a common condition in people with learning disability (LD), the risk of suffering from epilepsy increases with the severity of the LD[1] and some syndromes have LD and epilepsy as an integral part (e.g. Lennox–Gastaut). Point prevalence studies have revealed epilepsy rates of 40–50% in those with severe LD[1].

Historically the management of epilepsy in this group of people was the domain of the LD psychiatrist in his role as medical superintendent of the hospital[1]. Often these practitioners had no specialist training in epilepsy and this may have led to inappropriate management. With the closure of the large institutions and the move into the community the general practitioner manages epilepsy. With regard to psychotropic medication, such reviews are often conservative and clients continue on the same medication they were taking on discharge (pers. comm. B. Louis). Probably the same is true of anticonvulsants. However, the general practitioner has the choice of referring to a neurologist or psychiatrist in LD for advice on the management of epilepsy. Opinions vary on which speciality serves this population better[2,3].

At the same time as the closure of the mental handicap hospitals there have been the introduction of newer anticonvulsants as add-on therapy in treatment-resistant epilepsy, such as vigabatrin, lamotrigine and gabapentin. These drugs have different modes of action and side-effect profiles and are currently licensed as add-on treatments. Many of the LD population have poorly controlled epilepsy and thus would be candidates for these new anticonvulsants[4,5].

We report two cases of poorly controlled epilepsy in people with LD, discharged into the community from a large hospital, on two anticonvulsants, in whom addition of lamotrigine was successful.

CASE 1

BH is a 61-year-old lady with a mild learning disability of unknown aetiology. She lives with two other ladies in a staffed house in the community, following her discharge from a mental handicap hospital 5 years previously. She was referred to the LD psychiatrist because of 'behaviour problems'. These problem behaviours had been occurring for at least 16 years and consisted of intermittent episodes where she became mute, apathetic, withdrawn and refused to eat and drink. There was evidence of psychomotor retardation. Her eyelids flickered and she took up bizarre postures, for example crucifix. During these episodes she was doubly incontinent. These episodes lasted from 2

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hours to 4 days and on average occurred 4–6 times a month, most often at weekends.

At the age of 9 she was diagnosed as suffering from epilepsy (tonic–clonic and absence seizures) and was commenced on primidone and phenytoin. An EEG performed when she was 48 revealed multifocal discharges with photosensitivity. The primidone and phenytoin were replaced by sodium valproate and carbamazepine. At this time she suffered 2–4 tonic–clonic seizures per year and was also diagnosed and treated for hypothyroidism.

Aged 52 she suffered carbamazepine toxicity and the dosage was reduced. At 60 years her general practitioner commenced Sinemet 110 after making a clinical diagnosis of Parkinson's disease.

A clinical diagnosis of non-convulsive generalized status epilepticus was made. An EEG confirmed the potential for a seizure disorder probably both multifocal and primary generalized. Blood tests including thyroid function tests were all within normal limits.

Lamotrigine was introduced initially at 25 mg nocte and gradually increased to 100 mg twice daily. Over a period of 4 months the carbamazepine was reduced and stopped and the sodium valproate continued at 500 mg twice daily with the intention of reducing the dosage at a later date.

There was a reduction in frequency and duration of the withdrawn, mute episodes and at 4 months after commencing lamotrigine there were no further episodes.

A further increase in the lamotrigine dosage to 200 mg twice daily led to complaints by the patient of unsteadiness, dizziness and nausea. On examination she was ataxic and showed signs of cerebellar dysfunction. Reduction of the lamotrigine to 150 mg nocte produced a resolution of these symptoms. In the following six months there has only been one brief withdrawn episode.

CASE 2

JJ is a 29-year-old man with severe learning disability with cerebral palsy, quadriplegic, with no speech and completely dependent on carers. He was referred to the Learning Disability Service as his tonic–clonic seizure frequency was steadily increasing following his resettlement in a community home a year ago from a large mental handicap hospital. At the time of first assessment, JJ was drowsy, drooping on his wheelchair with little response to verbal commands. He was on sodium valproate 800 mg bd and phenytoin 125 mg and 100 mg daily, together with other medications for constipation. There were no old medical files available, thus the type of epilepsy and investigations were unknown.

With gradual reduction in sodium valproate and the introduction of lamotrigine, there was a dramatic improvement in his general well-being. He became more alert, happy, smiling, responding to his name by turning his head, his appetite also improved and the physiotherapist reported that the motor power in his limbs started improving.

Seizure frequency was minimal three months after the start of lamotrigine, but there was a slight increase in frequency following the cessation of sodium valproate after five months. However post-ictal recovery is more rapid. His general well-being continues to improve, and it was certainly a rewarding experience for his carers and parents who visit him regularly.

DISCUSSION

Lamotrigine was introduced into the UK in 1991. It is a triazine compound which acts by inhibiting glutamate release by blocking sodium channels[6]. Its protein binding is around 55%. It is metabolized in the liver, largely by glucuorinadation and half life is 25–30 hours[7].

It is licensed in the UK as add-on treatment of partial seizures and secondarily generalized tonic–clonic seizures not satisfactorily controlled with other anti-epileptic drugs. Recently the drug has been licensed for use in children over two years of age[8]. It may also be effective in other types of epilepsy, including absence, myoclonic, tonic, clonic and tonic–clonic seizures[7] and perhaps in Lennox–Gastaut Syndrome[9].

Side-effects of lamotrigine are small, although higher doses can cause headache, dizziness, diplopia, drowsiness, nausea and vomiting which usually resolve on reduction of dosage. A maculopapular rash is seen in 3% of patients which may lead to withdrawal of the drug[7]. This appears shortly after the introduction of the drug. A slow introduction of lamotrigine reduces the incidence of rash[4].

The main drug interaction is with sodium

valproate which inhibits the metabolism of lamotrigine hence increasing its half-life to 60 hours[5], thus lamotrigine toxicity may occur at a lower dose as illustrated in Case 1.

Non-convulsive status epilepticus (NCSE) makes up at least a quarter of all status epilepticus[10]. Symptoms include continuous state of altered consciousness, rhythmic shaking, clonic twitching, automatisms and myoclonic jerking of the limbs or face. Speech is also reduced[11]. Ictal phenomena may suggest the presence of a psychiatric illness[12].

Thus misdiagnosis of non-convulsive status epilepticus is not uncommon. In the first case this had led to the carers perceiving the person in a negative light. It had also led to a late presentation to the service.

The cases highlight the need for regular review of people with epilepsy and learning disability. This is to ensure optimal treatment in the context of innovations in epilepsy management. There is a risk of therapeutic nihilism in the management of people with epilepsy and learning disability. Inappropriate use of certain anticonvulsants may also impair cognition and therefore further handicap the person. In order to improve the quality of life of an individual with LD and epilepsy, regular and continuing assessment is required of the risks and benefits of anticonvulsant medication. Both the cases showed positive changes in quality of life with the improvement in their seizure management.

The mood stabilizing effects of carbamazepine and sodium valproate are widely known and lamotrigine has been reported anecdotally, to improve well-being. This might have had an effect in our cases[13]. It is not known if this is a result of improved seizure control or of other unknown mechanisms.

These two cases reveal the utility of lamotrigine in people with intratractable epilepsy and learning disability.

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