A structured, multi-disciplinary approach for the management of epilepsy for people with intellectual disabilities

Phil Tittensor et al examine the challenges of managing epilepsy in those with intellectual disabilities in the Midlands
Background
A learning disability or intellectual disability (ID) is broadly defined as a person who possesses a Full Scale Intellectual Quotient (IQ) below 70, encompassing developmental delay and impaired cognition social and adaptive skills [Ring, 2013]. Fisher et al [2014] state that epilepsy is a disease of the brain defined by any of the following: at least two unprovoked (or reflex) seizures occurring greater than 24 hours apart, one unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years, or the diagnosis of an epilepsy syndrome.

People with ID are more likely to develop epilepsy than the general population, with a mean incidence ranging from 22-26% and increasing with the severity of the learning disability [Robertson et al, 2015]. These people can have a range of comorbidities that differ depending on the presence and degree of brain damage. Additionally, people with ID and epilepsy have a higher likelihood of communication, psychiatric, behavioural and drug sensitivity problems that make the condition more difficult to treat compared to those with higher IQ [Royal College of Psychiatrists, 2017]. They require expert support from a range of professionals.

People with ID have a significantly shorter life expectancy – 23 years for men and 27 years for women, when compared to the general population [Learning Disabilities Mortality Review – LeDeR, 2019]. According to LeDeR [2019], 4% of deaths in people with ID were due to epilepsy, compared to less than 1% of the general population. The National Sentinel Audit of Epilepsy Deaths [Hanna et al, 2002] reported that almost half are preventable with optimal treatment. Although nearly 20 years old, this statistic has not significantly changed [Kerr et al. 2020]. In Cornwall, an integrated epilepsy and ID service has significantly reduced mortality figures, with one incident of sudden unexpected death in epilepsy (SUDEP), the most common reason for epilepsy mortality, since the service was commissioned in 2010 [Shankar et al, 2015; Shankar, Jory, 2020]. With a Cornish population of 538,000, Shankar and Jory [2020] state that the number of epilepsy related deaths for people with ID in the county would be expected to be between one and three per annum, demonstrating that a multi-disciplinary approach can make a positive contribution to safety for this vulnerable group of people.

Aim
People with epilepsy and ID in South Staffordshire, Wolverhampton and parts of the Black Country in the West Midlands have, historically, experienced differences in care, depending upon the catchment area of each of the three NHS Trusts (one acute and two community – appendix 1) providing epilepsy services in the locality. There is considerable geographical overlap between the Trusts, but the services roughly align with three Clinical Commissioning Groups [Wolverhampton, Cannock Chase and South East Staffordshire and the
Seisdon Peninsular] with a combined population of approximately 626,000, approximately 2,700 of whom will be adults with ID and epilepsy (Appendix 2). Some people have been managed by neurology, some by ID services, and some access a joint clinic run by clinicians from both specialties. We have wanted to ensure that there is an equity of service for all people with epilepsy and ID in the area, aiming for the gold standard of a specialist epilepsy team commissioned by the liaison of adult, learning disability and physical health commissioners, with all the necessary components of epilepsy and learning disability care across the lifespan, ensuring a biopsychosocial approach [Kachika and Shankar, 2018]. We believe that this will improve care and enhance safety by providing optimal treatment and management of people with epilepsy and ID.

The joint service follows evidence based practice. Integral to this is research and audit. The discrete departments comprising the joint service are research active. We aim to critically evaluate our service outcomes, participate locally and internationally [EpiNet, 2020] in efforts to identify and understand epilepsy related deaths, and to participate in multi-centre research projects, such as the UK ID and anti-epileptic drugs registry [Shankar et al, 2015].

Many people with severe or profound ID will require the support of professional care staff. Education of carers about epilepsy in the UK is patchy [Shankar et al, 2017], and guidelines provided by the Joint Epilepsy Council of the UK and Ireland (JEC) expired in 2016 [JEC, 2012]. The Epilepsy Nurses Association (ESNA), in conjunction with the International League Against Epilepsy (ILAE) and Royal College of Psychiatrists (RCPsych), published new guidelines recently [ESNA, 2019]. An important role for the joint service is to provide education and guidance for families, care staff and other health professionals, to raise the standards of care for people with epilepsy and ID, thereby improving safety and enhancing quality of life.

**Methods**

The Community Learning Disabilities Team (CLDT), and the Department of Neurology in mid and South Staffordshire (services currently provided by the Midlands Partnership NHS Foundation Trust and Royal Wolverhampton NHS Trust respectively), developed a joint epilepsy and intellectual disability service over 10 years ago. Originally conceived to improve the care of patients in an NHS residential facility, the service expanded, and now comprises bimonthly joint epilepsy and ID clinics run by a consultant nurse for the epilepsies and a consultant psychiatrist for people with ID. There is input into the clinic from nursing staff from the CLDT, who monitor patients between clinic visits, assist with medication changes and monitoring of seizures, and provide education and support to people with ID, their families and carers. This includes training in epilepsy awareness and midazolam administration in line with the latest guidelines [ESNA, 2019]. Patients have access to the full range of services offered by the CLDT, alongside the full range of

**Table 1**

**Interview regarding independence during the transitional period.**

<table>
<thead>
<tr>
<th>Change in seizure frequency</th>
<th>All seizure types (N=32)</th>
<th>Tonic Clonic seizures (N=24)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Seizure free</td>
<td>6%</td>
<td>8%</td>
</tr>
<tr>
<td>&gt;50% reduction</td>
<td>47%</td>
<td>29%</td>
</tr>
<tr>
<td>&lt;50% reduction</td>
<td>6%</td>
<td>8%</td>
</tr>
<tr>
<td>No change</td>
<td>19%</td>
<td>42%</td>
</tr>
<tr>
<td>Seizures worse</td>
<td>22%</td>
<td>17%</td>
</tr>
</tbody>
</table>
diagnostic facilities offered by the Department of Neurology. There are facilities for further neurological opinion and formal tertiary referral to the regional epilepsy centre. In 2019, we developed a similar service for people from Wolverhampton in collaboration with the Black Country Partnership NHS Trust CLDT.

The service advocates the use of EpsMon [SUDEP Action, 2020], an app to estimate an individual’s risk from seizures. This can be used by the family or care team to provide an early warning of deteriorating seizure control (the primary risk factor for SUDEP), enabling them to seek specialist medical advice.

**Results**

Clinics for the last five years have been analysed. In that time, 33 people have accessed the service. 18 remain under ongoing review. Of those people discharged from the joint service, five have continued to be seen by the learning disability service, three have been discharged to their GP with no ongoing issues requiring specialist input (all patients have the opportunity for carers to self-refer back to the epilepsy service in the event of problems), two have moved out of the area, one discharged to neurology only follow-up and four have died (no deaths were epilepsy-related).

Over the five years, there have been 158 consultations in 25 joint clinics. About a third of patients had mild ID (30%), a third moderate ID (30%), and a third severe or profound ID (40%). All the patients had at least one comorbidity in addition to their ID and epilepsy, the most common being cerebral palsy (29%), autism (19%), genetic syndromes (19%) and other physical conditions (e.g. diabetes, cardiac conditions) (52%). 97% of the patients had involvement from the CLDT, and 87% had documented, up to date, risk assessments and epilepsy care plans in place. 86% of the patients had a documented epilepsy syndrome or classification using the current ILAE system [Fisher et al, 2017]. 95% of patients using the service have documented EEG, but only 43% had inter-cranial imaging (MRI or CT). 45% of patients were taking two anti-epileptic drugs, with 15% on monotherapy. Conversely, 9% were taking four or more anti-epileptic drugs. 70% had never tried one of the newer anti-epileptic drugs*. The seizure outcomes of patients using the joint epilepsy and ID service are summarised in Table 1.

**Discussion**

The joint epilepsy and ID clinic supports a relatively small but extremely complex group of people. Their epilepsy is highly refractory, with the majority of patients requiring more than one anti-
Epileptic drug to manage the condition. All have comorbidities that can impact upon the choice of anti-epileptic drug treatment and epilepsy itself. Despite this, over half of the patients accessing the service have seen a greater than 50% reduction in their seizures, with roughly double the seizure freedom rate that might be expected from a highly refractory cohort [Kwan and Brodie, 2000]. Given that improvements in seizure control is one of the biggest factors influencing SUDEP Hanna et al, 2002], the audit results indicate that patients may have a reduced mortality risk since they accessed the service. There is further evidence for this with the widespread use of risk assessments, epilepsy care plans and the involvement of the CLDT. We are currently exploring methods of identifying all epilepsy related deaths in Wolverhampton and South Staffordshire, in conjunction with the local coroner, in order to better understand the risks locally, as well as the impact of the joint service.

Fewer patients have accessed inter-cranial imaging than might be expected. There are a number of reasons for this, the most common being the difficulties in compliance with the requirements of the examination, particularly in the case of MRI where the patient needs to lie still for around half an hour. There was also a surprisingly low number of patients who had tried one of the newer anti-epileptic drugs, which we defined as those receiving marketing licences in the UK since 2006*. Anecdotally, many professional and family carers have voiced concerns about mood changing side-effects associated with some of the newer drugs, which could account for their relatively low use.

The present service offers support to a highly selective group of patients with difficulties relating to their ID as well as their epilepsy. There would need to be a considerable increase in provision to offer this joint approach to all individuals with ID and epilepsy in our area. However, the service is undergoing rapid expansion, with another epilepsy specialist nurse, and several consultant psychiatrists and specialist nurses for people with ID interested in the approach.

Further service evaluation and audit are planned in order to ensure that we place patients, their families and carers at the heart of the service. This focus on quality will help us to identify the benefits of the joint approach as well as any problems. Continual audit and evaluation will also guide us when considering further development of the service.

While the majority with ID and epilepsy will be well managed in either service, there are a number who need the expertise of both services.

Conclusion
The management of epilepsy in people with intellectual disability is often challenging and complex. Seizure freedom is more difficult to achieve in this population, and in the long-term, specialist support is likely to be needed. While the majority of people with ID and epilepsy will be well managed in either neurology or ID services, there are a number who need the expertise of both services. Given the significant interplay between comorbidities commonly associated with intellectual disability and epilepsy, we believe that best practice is to develop joint clinics and close working relationships between the CLDT and Department of Neurology. Further study is required to examine outcomes for people with intellectual disability served by neurology services alone, intellectual disability services alone and a joint service such as that which is described in this article.

*Zonisamide, Pregablin, Rufinamide, Lacosamide, Eslicarbazepine, Retigabine, Perampanel, Brivaracetam.

Tittensor PA, Rowe J, Youssef C, Manfredonia F: The Royal Wolverhampton NHS Trust
Baker S: Midlands Partnership NHS Foundation Trust
Varghese S: Black Country Health NHS Foundation Trust

Appendix 1. Participating Trusts.
The Royal Wolverhampton NHS Trust
Midlands Partnership NHS Foundation Trust
Black Country Health NHS Foundation Trust

Appendix 2. Local epidemiology.
The three Clinical Commissioning Groups covered by our services (Wolverhampton, Cannock Chase and South East Staffordshire and the Seisdon Peninsular) have a combined population of approximately 626,000. 2.4% of people in the UK have ID [Mental Health Foundation, 2011], so there are approximately 15,000 people with ID in the area. Using Robertson et al’s [2015] incidence estimates, the local population of people with epilepsy and ID is about 3,600. 25% will be children, so there are about 2,700 adults with ID and epilepsy in the area.
Further reading

Epilepsy Nurses Association (ESNA) (2019). Best practice guidelines for training professional carers in the administration of buccal (or mucosal) midazolam for the treatment of prolonged and/or clusters of epileptic seizures in the community. www.esna-online.org (accessed 22/04/2020).


