Abstract
Epilepsy is more prevalent in people with learning disabilities than in the general population and there are a number of rare epilepsy syndromes closely associated with learning disability. These syndromes are challenging to manage, notably because seizures are often refractory, the risk of status epilepticus is high and comorbidities are usually present. People with a rare epilepsy syndrome need high levels of specialist support throughout their lives. Such syndromes are rare, but professionals who care for people with learning disabilities are likely to encounter them at some point in their career. Improved knowledge and understanding of rare epilepsy syndromes can assist learning disability nurses to provide adequate information, care and support to service users, family members and paid carers as well as to other healthcare professionals.

This article explains the association between epilepsy and learning disability, describes rare epilepsy syndromes and explores the goals of treatment, additional treatment options and challenges of management.

Aim and intended learning outcomes
The aims of this article are: to enhance learning disability nurses’ knowledge of rare epilepsy syndromes and their relationship with learning disability; to help nurses care for people with a rare epilepsy syndrome who also have a learning disability; and to help nurses provide support and education to family carers, paid carers and other healthcare professionals. After reading this article and completing the time out activities you should be able to:

- Provide examples of syndromes where epilepsy and learning disability typically co-exist.
- Describe typical features of epilepsy in people with learning disabilities.
- Explain the goals of treatment in rare epilepsy syndromes.
- Outline additional treatment options for improving seizure control and optimising quality of life.
- Detail the comorbidities usually associated with rare epilepsy syndromes.
TIME OUT 1
Has your team cared for a child, young person or adult who had a rare epilepsy syndrome? Can you recall the syndrome's name, features and treatment? Can you describe the challenges it posed for the person and those caring for them?

Introduction
Epilepsy is a disorder of the brain characterised by recurrent seizures. It is one of the most common neurological disorders, affecting about 50 million people worldwide (World Health Organization 2023). In 50% of people with epilepsy the underlying cause is unknown. Epilepsy is thought to encompass more than 25 different syndromes (Habibi et al 2016), a syndrome being a collection of features that usually occur together. The features of epilepsy syndromes include, for example, the type of seizures commonly seen, the age at which seizures commonly start, the usual course of seizures, the part of the brain involved and the underlying genetic characteristics (Epilepsy Foundation 2013).

Epilepsy can affect anyone but is particularly common among people with learning disabilities. The prevalence of epilepsy is estimated to range between 14% and 44% in people with learning disabilities (Bowley and Kerr 2000, McCarron et al 2014) and between 0.6% and 1% in people without learning disabilities (Forsgren et al 2005, Linehan et al 2010, Ngugi et al 2010). The more severe the learning disability, the higher the risk of epilepsy (Robertson et al 2015). Epilepsy can develop at any age but epilepsy in people with learning disabilities often starts in early childhood (Ryan et al 2021).

There are a number of learning disability syndromes in which epilepsy is common. There are also epilepsy syndromes in which learning disability is highly prevalent. These epilepsy syndromes are rare but, despite being rare, they are collectively significant in the sense that they affect an important proportion of people receiving care and treatment for epilepsy (Balestrini et al 2021). Because of the high prevalence of epilepsy among people with learning disabilities, healthcare professionals who work with this group are likely to encounter these rare epilepsy syndromes at some point in the course of their career.

TIME OUT 2
You may have heard about status epilepticus and be aware that it is a medical emergency, but would you be able to describe it accurately to a junior nurse, nursing student or family carer? Think about what you would tell them. You can find information and inspiration in this podcast: https://www.youtube.com/watch?v=lyLmEWYSz4Q

Rare epilepsy syndromes
Epilepsy is considered rare when its underlying cause affects fewer than 200,000 people (Rare Epilepsy Network (REN) 2023). The incidence of rare epilepsy syndromes is largely unknown (Tian et al 2020). Rare epilepsy syndromes often have an early onset (Scheffer et al 2017). Their diagnosis can take time and they can be misdiagnosed, but with improved genetic testing, screening, imaging and encephalography, they tend to be diagnosed quicker than in the past (REN 2023).

Rare epilepsy syndromes can be medically complex (REN 2023) and are often challenging to treat (Scheffer et al 2017). There is a high prevalence of comorbidities, which compounds the difficulties of management (Ho et al 2018). People with a rare epilepsy syndrome often have a learning disability (Tschamper and Systad 2022), the presence of which is likely to compound communication difficulties and make it more challenging for healthcare professionals to recognise and understand the person’s symptoms and needs (Robertson et al 2015). Rare epilepsy syndromes where learning disability is present have unique presentations and comorbidities. They require a high level of expert support and care from a multidisciplinary team (Balestrini et al 2021).
The most severe and complex epilepsy syndromes include early-onset developmental and epileptic encephalopathies, which are characterised by refractory seizures, the presence of a learning disability and other comorbidities, and usually a genetic aetiology (Balestrini et al 2021). In some epilepsy syndromes, electroencephalographic features and seizure types evolve over time (Cross et al 2017), so presentation and symptoms may change (Both et al 2018). The various comorbidities associated with rare epilepsy syndromes may also change over time (REN 2023).

People with a rare epilepsy syndrome are likely to experience frequent episodes of status epilepticus (SE), a life-threatening neurological condition defined as a prolonged seizure lasting for five minutes or longer or recurrent seizures one after the other without recovery in between (National Institute for Health and Care Excellence (NICE) 2023). Mortality due to SE is higher in people with a rare epilepsy syndrome than in people with epilepsy in general; for example, one third of deaths in people with Dravet syndrome are attributed to SE (Devinsky et al 2015).

Epilepsy in people with learning disabilities can have negative effects on their activities of daily living and quality of life, increasing their care needs and risk of death (McCarron et al 2014, Shankar et al 2018). Long-term outcomes for people with a rare epilepsy syndrome are considered to be poor (Asadi-Pooya 2018, Balestrini et al 2021). Data on survival and outcomes in adulthood are limited for the more complex epilepsy syndromes such as those with early onset in childhood (Balestrini et al 2021).

Box 1 describes five syndromes where epilepsy and learning disability typically co-exist – but there are others.

Diagnosing a specific epilepsy syndrome, describing its features and determining its underlying cause is important to inform decision-making and guide management, since it provides crucial information on which treatments may benefit the person (Epilepsy Foundation 2013, Symonds et al 2021, REN2023). It is therefore crucial to investigate potential epilepsy syndromes in people with learning disabilities if it is suspected that they may have one, and if an epilepsy syndrome is present, to identify its underlying cause (Ryan et al 2021, Symonds et al 2021).

**TIME OUT 3**

How well do you know the syndromes described in Box 1? Use this ‘time out’ to search for more information about all, some or one of these syndromes, according to your interests. You could look up these websites, for example:

- www.angelmanuk.org/what-is-angelman-syndrome
- www.epilepsy.org.uk/info/syndromes/dra\vet-syndrome
- www.epilepsy.org.uk/info/syndromes/lennox-gastaut-syndrome
- www.rettuk.org/what-is-rett-syndrome
- www.tuberous-sclerosis.org/information-and-support/what-is-tsc

**Goals of treatment**

The treatment of epilepsy syndromes is often complex (Balestrini et al 2021). In epilepsy syndromes, seizures are often numerous in type and frequency, and total freedom from seizures is elusive. According to the Epilepsy Foundation (2013), only 5% to 10% of people with refractory epilepsy achieve full seizure control even when trialling different anti-seizure medicines or increasing their number. Furthermore, people with a rare epilepsy syndrome are usually at increased risk of SE compared with people with epilepsy in general (Gibson 2014, Dravet 2016). As a consequence, treatment should not focus on full seizure control but on:

» Improving seizure control.
» Optimising quality of life.
» Preventing or treating SE.

**Improving seizure control**

In their respective articles on Lennox-Gastaut syndrome and Dravet syndrome, Asadi Pooya (2018) and Strzelczyk and Schubert-Bast (2022) noted that broad-
spectrum anti-seizure medicines are generally insufficient to fully control seizures and that the goal of treatment should therefore be to reduce the frequency of the most incapacitating seizures. This would, in turn, reduce hospital admissions for complications such as prolonged seizures and seizure-related injuries. The risk of sustaining an injury during a seizure, for example a fracture, is high and seizure-related injuries are a common cause of presentation to hospital (Morton-Nance 2022).

Optimising quality of life
In people with a rare epilepsy syndrome, improving seizure control is crucial for optimising health-related quality of life (Bjurulf et al 2022a, 2022b). Optimising quality of life should be one of the goals of treatment because long-term outcomes are generally poor (Asadi-Pooya 2018) and treatment is unlikely to lead to full seizure control (Cross et al 2017). Important factors to consider are the potential adverse effects of medicines and drug interactions, since these may reduce the person’s quality of life and exacerbate comorbidities (Cross et al 2017).

Preventing or treating status epilepticus
SE is a medical emergency requiring prompt recognition and treatment. Benzodiazepines are the first-line treatment option for SE in the community (Ryan et al 2021). Prolonged seizures leading to SE are a major concern in people with a rare epilepsy syndrome, who commonly require the administration of rescue medicines, usually buccal (oromucosal) midazolam, to prevent a prolonged seizure developing into SE or to treat SE (Bjurulf et al 2022a, 2022b).

Additional treatment options
There are treatment options that can be used in addition to medicines to improve seizure control and optimise people’s quality of life. Table 1 gives an overview of four such options: ketogenic diet, vagus nerve stimulation (VNS), cannabidiol (CBD)-enriched products and surgery.

Box 1. Five syndromes where epilepsy and learning disability typically co-exist

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Angelman syndrome</td>
<td>Rare neurodevelopmental disorder characterised by developmental delay, learning disability, seizures, communication difficulties, characteristic behaviours and movement disorders (Duis et al 2023)</td>
</tr>
<tr>
<td>Dravet syndrome</td>
<td>Severe developmental and epileptic encephalopathy (Scheffer et al 2017) with tonic-clonic seizures starting in the first year of life and evolving into status epilepticus (SE) and myoclonic and atypical absences</td>
</tr>
<tr>
<td>Lennox-Gastaut syndrome</td>
<td>Rare epilepsy syndrome and severe form of epilepsy that emerges in childhood and is characterised by tonic seizures, atypical absences and atonic drop attacks (Asadi-Pooya 2018)</td>
</tr>
<tr>
<td>Rett syndrome</td>
<td>Severe neurodevelopmental disorder primarily affecting girls, with an initial period of normal development followed by a regression of acquired skills between the ages of six months and 12 months (Hagberg et al 2002)</td>
</tr>
<tr>
<td>Tuberous sclerosis</td>
<td>Also known as tuberous sclerosis complex</td>
</tr>
</tbody>
</table>

TIME OUT 4
Think back to a person with a rare epilepsy syndrome who your team supported in the past. Did the person have an individualised care plan? What treatments did they receive and what were the goals of treatment? Did the treatment have an effect on seizure control and quality of life? Did the person and their family have realistic expectations of treatment?
### Challenges of management

**Collaborative care**

People with a rare epilepsy syndrome need high levels of support throughout their lives, starting in early childhood when seizures typically commence, from family and/or paid carers and from healthcare professionals.

Individualised care and support from a multidisciplinary team that includes specialist clinicians is crucial. Carers need to be considered as part of the care team, since they have insider knowledge of the person, what their condition means for them and what their needs are (Both et al 2018).

---

**Table 1. Overview of four additional treatment options**

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Procedure and benefits</th>
<th>Considerations for clinical practice</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ketogenic diet</td>
<td>» Dietary therapy high in fat and low in carbohydrates and protein</td>
<td>» Requires exact calculations of quantities of food from different food groups and adherence to specific quantities of food and recipes</td>
</tr>
<tr>
<td></td>
<td>» Most well-known dietary therapy used to treat epilepsy, with evidence that people experience fewer seizures (Epilepsy Foundation 2017)</td>
<td>» Must be approved by the person’s neurologist or epileptologist and overseen by the multidisciplinary team</td>
</tr>
<tr>
<td></td>
<td>» Recommended in people with a rare epilepsy syndrome and identified as a tolerated and effective option in people with Lennox-Gastaut syndrome (Asadi-Pooya 2018)</td>
<td>» The person and their carers must understand what following the diet requires and what they can expect from it</td>
</tr>
<tr>
<td>Vagus nerve stimulation (VNS)</td>
<td>» A small device implanted in the upper left side of the chest sends regular electric impulses to the brain, via a thin wire that connects it to the vagus nerve in the neck, in the hope that it will disrupt the electrical signals in areas of the brain where seizures occur</td>
<td>» Implanting the VSN device requires pre-surgical assessment and testing and a small surgical operation</td>
</tr>
<tr>
<td></td>
<td>» The VSN device comes with a separate magnet that can be swiped across the chest where the device is implanted as part of seizure first aid</td>
<td>» The person may need tailored support before and during surgery such as preparatory work, familiarisation, social stories and reasonable adjustments</td>
</tr>
<tr>
<td></td>
<td>» VNS can support a reduction in seizures (Asadi-Pooya 2018) but must be used in conjunction with anti-seizure medicines</td>
<td>» Some people may not tolerate pre-surgical testing and some may have comorbidities that make it challenging for them to undergo surgery</td>
</tr>
<tr>
<td></td>
<td>» The majority of people who receive VNS report improved quality of life (Epilepsy Foundation 2018)</td>
<td>» VNS does not cure epilepsy and it does not work for everyone. Although the person may feel better in themselves, there may be no reduction in seizures (Epilepsy Foundation 2018). It is important that the person and their carers have realistic expectations of treatment</td>
</tr>
<tr>
<td>Cannabidiol (CBD)-enriched products (Epidiolex or Epidyolex)</td>
<td>» Emerging interest in the use of CBD-enriched products for treating drug-resistant epilepsy (Golub and Reddy 2021)</td>
<td>» CBD-enriched products are licensed for use from the age of two years onwards in Dravet syndrome, Lennox-Gastaut syndrome and tuberous sclerosis. In Dravet syndrome and Lennox-Gastaut syndrome, CBD-enriched products are licensed for use in conjunction with the anti-seizure medicine clobazam (Epilepsy Action 2023, Epilepsy Ireland 2023)</td>
</tr>
<tr>
<td></td>
<td>» Evidence from randomised, double-blind, placebo-controlled trials has led, in the US and European countries including in the UK, to the approval of CBD-enriched products for Dravet syndrome, Lennox-Gastaut syndrome and tuberous sclerosis; for example, Devinsky et al (2017) demonstrated a reduction in seizures in 43% and seizure freedom in 5% of participants with Dravet syndrome</td>
<td>» Treatment with CBD-enriched products needs to be monitored to determine its efficacy in reducing seizures. The person needs to be observed for adverse reactions and side effects. Education about, and adherence to, treatment are required</td>
</tr>
<tr>
<td></td>
<td>» Adjunct CBD-enriched products have been found to be generally safe and effective for treatment-resistant seizures in children with severe early-onset epilepsy, but their efficacy in the long-term treatment of various epilepsy and seizure types in adults is still being tested (Golub and Reddy 2021)</td>
<td></td>
</tr>
<tr>
<td>Surgery</td>
<td>» A treatment option for people with certain types of refractory epilepsy for many years</td>
<td>» The person must be able to tolerate the required pre-surgical investigations and assessments, which include video encephalography with sleep recording, magnetic resonance imaging and neuropsychological assessment (Asadi-Pooya et al 2021)</td>
</tr>
<tr>
<td></td>
<td>» Surgical procedures include resection, corpus callosotomy (which consists in separating the bundle of fibres connecting the two brain hemispheres) and deep brain stimulation (Jackson et al 2015). Which procedure is chosen depends on factors such as seizure type, area of brain affected and localisation of epileptogenic regions</td>
<td></td>
</tr>
<tr>
<td></td>
<td>» Surgery usually aims to reduce the occurrence of the most incapacitating seizures, such as tonic-clonic seizures. Among people with Lennox-Gastaut syndrome who underwent surgery, almost 39% were free of these types of seizures one year after surgery (Asadi-Pooya et al 2013, Asadi-Pooya 2018)</td>
<td></td>
</tr>
</tbody>
</table>
The involvement of specialists with expertise in epilepsy and specialists with expertise in learning disability is also crucial. This is because non-specialists may lack the skills required to communicate with the person and navigate the difficulties of management, including decision-making when people may lack mental capacity to make decisions for themselves (Balestrini et al 2021). Furthermore, non-specialists may not have sufficient understanding of epilepsy and rare epilepsy syndromes – for example, the fact that the person’s condition and symptoms may evolve over time (Cross et al 2017).

Every person needs an individualised care plan tailored to their needs and lifestyle. The care plan needs to be agreed between the person, their carers and the professionals looking after them (Ryan et al 2021). It should be focused on optimising quality of life through improved seizure control, on monitoring the adverse effects of treatment and on responding efficiently to prolonged seizures and SE. The care plan should outline the goals of treatment and include a rescue medicines protocol. It is essential that it is adhered to (Both et al 2018).

Comorbidities
Comorbidities in people with a rare epilepsy syndrome can affect their condition and compound the difficulties of management, thereby severely reducing their quality of life and that of their carers. As the person becomes older, the burden of comorbidities can surpass that of seizures (Marchese et al 2021). Beyond the treatment of the epilepsy itself, the assessment and management of comorbidities should therefore be a core aspect of care. Everyone who cares for a person with a rare epilepsy syndrome, professionally or non-professionally, needs to receive education about comorbidities and be wary of them and their effects on the person (Devinsky et al 2015, Cross et al 2017, Marchese et al 2021).

Comorbidities associated with rare epilepsy syndromes include:

» Cognitive impairment – moderate-to-severe cognitive impairment is common in people with a rare epilepsy syndrome (Marchese et al 2021).

» Mental health issues – for example depression, anxiety and psychiatric disorders (Marchese et al 2021, Mula et al 2021).

» Behavioural issues – which can include agitation, aggression, hyperactivity, autism traits and social issues (Ho et al 2018).

» Sleep disturbances – sleep disturbances are common in people with a rare epilepsy syndrome and can compound the cognitive, mental health and behavioural issues outlined above (Marchese et al 2021, Van Nuland et al 2021). Sleep disturbances and sleep deprivation can occur due to nocturnal seizures and post-seizure recovery.

» Mobility impairment – mobility can become impaired due to repetitive and aggressive seizures such as atonic seizures, which can lead to various levels of injury (Kerr et al 2011). All motor domains including balance, coordination and power of movement can be negatively affected (Verheyen et al 2019).

Information and education
Healthcare professionals and carers need information and education so they can work collaboratively to manage the person’s condition, enhance their quality of life and reduce morbidity and mortality (O’Dwyer et al 2018). It is essential that everyone who cares for a person with a rare epilepsy syndrome, professionally or non-professionally, knows how to safely support them during a seizure and how to administer rescue medicines to prevent or treat SE (Both et al 2018). Carers require specific information and education to help them in their role. Tschamper and Systad (2022) conducted a scoping review on the self-reported information needs of formal and informal carers of people with a rare epilepsy syndrome.
syndrome. Carers needed medical information on how to prevent seizures and how to administer rescue medicines. If carers are not given clear disease-specific information, the consequences for the person’s health can be extremely serious (Tschamper and Systad 2022). Beyond medical information, carers also need information on how to cope with emotional distress, experiential information from peers and interdisciplinary information exchange (Tschamper and Systad 2022).

Transition to adult services
Balestrini et al (2021) emphasised the importance of support from carers throughout the person’s life, especially in the phase of transition from children’s to adult services. That transition can be extremely challenging (Kerr et al 2011). In people with a rare epilepsy syndrome, characteristics usually seen in childhood, such as the types of seizure experienced, are often maintained in adolescence and adulthood. This increases the likelihood of inadequate service provision once the person has transitioned to adult services because professionals in adult services may not have come across these characteristics in adults. The term ‘childhood epilepsies’ refers to epilepsies with an onset in childhood, but it can be misunderstood as referring to epilepsies that only affect children, whereas the epilepsies usually continue in adolescence and adulthood (Balestrini et al 2021).

The challenges associated with the transition to adult services partly stem from how these services are conceived and delivered. In adult services, the onus is usually on the person to understand their condition and ask for support when needed, follow-up visits are less frequent (Camfield et al 2012) and support is more primary care-based, with greater involvement of the person’s GP (Balestrini et al 2021). This is a major change compared with children’s services and can adversely affect the care of young people and adults (Balestrini et al 2021).

Learning disability nurse role
The role of learning disability nurses in supporting people with a rare epilepsy syndrome and a learning disability, and their carers, is paramount. In some complex cases, the absence of a learning disability nurse can negatively affect the quality of care and the person’s outcomes (Morton-Nance 2022).

Learning disability nurses have a role in the administration of rescue medicines – usually buccal midazolam – and in ensuring all carers are familiar with their administration, including dose, timing, route, technique and observation of potential adverse effects such as respiratory depression (Ryan et al 2021). Some nurses may also be involved in prescribing these medicines.

Another role of learning disability nurses is educating others involved in the person’s care, whether in a professional or a non-professional role, who may become discouraged by the fact that seizures are ongoing. Nurses can help them understand the person’s condition and its presentation, how it is likely to evolve, that seizures may be refractory and can lead to SE, what the treatment options are and how to manage comorbidities.

Nurses can also remind them of the goals of treatment outlined in the person’s care plan. NICE (2022) recommends repeating information as needed, providing a copy of the care plan to service users and carers, and giving them the opportunity to discuss their concerns.

Learning disability nurses’ role further includes providing information about epilepsy to anyone who comes into contact with children, young people and adults with epilepsy and in educating the wider public about epilepsy to reduce the stigma associated with it – the existence of such stigma is confirmed in Kwon et al (2022). Nurses need to know the existing guidance and resources, such as the NICE (2022) guideline on epilepsies, and keep up to date with best practice, so that they can lead the care team in addressing people’s
needs across the lifespan through optimal care delivery and liaison between services (Doody et al 2023).

**Conclusion**

There are a number of epilepsy syndromes in which learning disability is typically present. These syndromes are rare but learning disability nurses are likely to come across them at some point in their professional life. All those who care for people with a rare epilepsy syndrome, whether in a professional or non-professional role, need to understand the person’s condition and its manifestations, likely evolution, comorbidities and treatment options. Treatment should focus on improving seizure control and optimising quality of life while ensuring SE is efficiently prevented or treated. The person will need lifelong support from a care team that includes specialist clinicians and the person’s family and/or paid carers. The care team will need disease-specific information and education so that they can work collaboratively to treat the epilepsy, manage the comorbidities and optimise the person’s quality of life, particularly during the transition from children’s to adult services.

**TIME OUT 5**

Identify how understanding treatment goals and management challenges in rare epilepsy syndromes applies to your practice and the requirements of your regulatory body

**TIME OUT 6**

Now that you have completed the article, reflect on your practice in this area and consider writing a reflective account. See: rcni.com/reflective-account

---

**References**


Dravet C (2016) Dravet syndrome: addressing the needs of patients and families: introduction. Canadian Journal of Neurological Sciences. 43, Suppl3, S1-S2. doi:10.1017/cjn.2016.64


Dravet Foundation (2023) What is Dravet Syndrome? dravetfoundation.org/what-is-dravet-syndrome (Last accessed: 20 November 2023.)


Epilepsy Ireland (2023) Evidence in Epilepsy. www.epilepsy.ie/content/evidence-epilepsy (Last accessed: 20 November 2023.)


---

See: rcni.com/reflective-account
Rare Epilepsy Network (2023) What is a Rare Epilepsy? www.rareepilepsynetwork.org/what-are-rare-epilepsies (Last accessed: 20 November 2023.)
World Health Organization (2023) Epilepsy. www.who.int/news-room/fact-sheets/detail/epilepsy
Rare epilepsy syndromes

1. Which of the following statements about epilepsy syndromes is accurate?
   a) They are rare types of epilepsy with early onset in childhood that usually resolve before adulthood
   b) They are among the most common neurological disorders, affecting about 50 million people worldwide
   c) Their features include the age at which seizures start, the usual course of seizures and the part of the brain involved
   d) They are early-onset developmental and epileptic encephalopathies

2. Which of these statements about the relationship between epilepsy and learning disability is inaccurate?
   a) Epilepsy can affect anyone but is particularly common among people with learning disabilities
   b) There are epilepsy syndromes in which learning disability is highly prevalent
   c) The less severe the learning disability, the higher the risk of epilepsy
   d) Epilepsy can develop at any age but epilepsy in people with learning disabilities often starts in early childhood

3. The incidence of rare epilepsy syndromes is:
   a) Largely unknown
   b) Common knowledge
   c) Often discussed in the medical literature
   d) Open to interpretation

4. Early-onset developmental and epileptic encephalopathies are characterised by which of the following?
   a) Refractory seizures
   b) The presence of a learning disability
   c) The presence of other comorbidities
   d) All of the above

5. What is the definition of status epilepticus (SE)?
   a) A prolonged seizure lasting at least 15 minutes or recurrent seizures without recovery in between
   b) A prolonged seizure lasting at least five minutes or recurrent seizures with phases of recovery in between
   c) A seizure from which the person is unlikely to recover
   d) A prolonged seizure lasting at least five minutes or recurrent seizures without recovery in between

6. What should not be a focus of treatment in people with a rare epilepsy syndrome?
   a) Full seizure control
   b) Improving seizure control
   c) Optimising quality of life
   d) Preventing or treating SE

7. Which of the following is an additional treatment option to improve seizure control?
   a) Mediterranean diet
   b) Recreational cannabis
   c) Vagus nerve stimulation
   d) Cosmetic surgery

8. It is important that the person's individualised care plan:
   a) Focuses exclusively on controlling seizures with medicines
   b) Is agreed between them, their carers and professionals
   c) Clarifies that the main treatment goal is full seizure control
   d) Clarifies why a rescue medicines protocol is not needed

9. Which of the following is not a comorbidity always associated with rare epilepsy syndromes?
   a) Cognitive impairment
   b) Obstructive sleep apnoea
   c) Depression
   d) Impaired balance and coordination

10. Nurses can help carers of people with a rare epilepsy syndrome and learning disability to:
    a) Understand the person’s condition and its presentation
    b) Understand that seizures may be refractory and can lead to SE
    c) Understand the treatment options and how to manage comorbidities
    d) All of the above

How to complete this assessment

This multiple-choice quiz will help you test your knowledge. It comprises ten multiple-choice questions broadly linked to the previous article. There is one correct answer to each question.

You can read the article before answering the questions or attempt the questions first, then read the article and see if you would answer them differently.

You may want to write a reflective account. Find out how at rcni.com/reflective-account

You can go online to complete this multiple-choice quiz at rcni.com/cpd/test-your-knowledge and save it to your RCNi portfolio to help meet your revalidation requirements.

This multiple-choice quiz was compiled by Anne-Claire Bouzanne

The answers to this quiz are: 1. c 2. c 3. a 4. d 5. d 6. a 7. c 8. b 9. b 10. d

You’ve read the article, now test your knowledge

As a subscriber, you have access to a library of interactive ‘Test your knowledge’ quizzes. Each quiz is based on a peer-reviewed article and consists of ten multiple-choice questions.

Complete a quiz, save it instantly to your RCNi Portfolio, and quickly build your personal CPD hours for revalidation. You can even write and save a reflective account on what you’ve just learnt.