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The epilepsies

Support for education and learning: clinical case scenarios - Children and young people with epilepsy

February 2012

NICE clinical guideline 137

These clinical case scenarios accompany the clinical guideline: ' The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care' (available at www.nice.org.uk/guidance/CG137).

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Introduction

NICE clinical case scenarios

Clinical case scenarios are an educational resource that can be used for individual or group learning. Each question should be considered by the individual or group before referring to the answers.

These four clinical case scenarios have been put together to improve your knowledge of the epilepsies and its application in practice. They illustrate how the recommendations from 'the epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care', (NICE clinical guideline 137 http://guidance.nice.org.uk/CG137) can be applied to the care of children and young people presenting within primary care.

The clinical case scenarios are available in two formats: this PDF, which can be used for individual learning, and a slide set that can be used for groups. Slides from the clinical case scenario slide set can be added to the standard NICE slide set (http://guidance.nice.org.uk/CG137/SlideSet/ppt/English) produced for this guideline.

You will need to refer to the NICE clinical guideline to help you decide what steps you would need to follow to diagnose and manage each case, so make sure that users have access to a copy (either online at www.nice.org.uk/guidance/CG137 or as a printout). You may also want to refer to the epilepsy NICE pathway (http://pathways.nice.org.uk/pathways/epilepsy) and the specialist library page on NHS Evidence (https://www.evidence.nhs.uk/topic/epilepsies?q=epilepsy).

Each case scenario includes details of the child or young person's initial presentation. The clinical decisions about diagnosis and management are then examined using a question and answer approach. Relevant recommendations from the NICE guideline are quoted in the text (after the answer), with corresponding recommendation numbers.

The epilepsies

Epilepsy is a common neurological disorder characterised by recurring seizures. Different types of epilepsy have different causes. Accurate estimates of incidence and prevalence are difficult to achieve because identifying people who may have epilepsy is difficult. Epilepsy has been estimated to affect between 362,000 and 415,000 people in England. In addition, there will be further individuals, estimated to be 5–30%, so amounting to up to another 124,500 people, who have been diagnosed with epilepsy, but in whom the diagnosis is incorrect. Incidence is estimated to be 50 per 100,000 per year and the prevalence of active epilepsy in the UK is estimated to be 5–10 cases per 1000. Two-thirds of people with active epilepsy have their epilepsy controlled satisfactorily with anti-epileptic drugs (AEDs). Other approaches may include surgery. Optimal management improves health outcomes and can also help to minimise other, often detrimental, impacts on social, educational and employment activity. ‘The epilepsies’ (NICE clinical guideline 20) stated that the annual estimated cost of established epilepsies was £2 billion (direct and indirect costs).

Clinical case scenarios for children and young people

Case scenario 1: Leanne

Presentation

Leanne is a 6-year-old girl whose teachers have suggested that her parents take to her GP. They have noticed that she seems to have problems listening and to be daydreaming a lot in class.

The GP asks whether her parents have also seen her daydream. Her mother has, but has not thought much about it. However, more recently, it seems to have been happening more frequently. On direct questioning by the GP, Leanne's mother thinks that these daydreams or ‘trances’ as she calls them sometimes occur when Leanne is in the middle of doing or saying something, and they interrupt her activity.

Leanne’s birth and early medical history, including her development, have been normal. There was a history of epilepsy on her father’s side of the family. Her younger brother and older sister are well.

1.1 Question

What should Leanne's GP consider as a possible diagnosis?

1.1 Answer

The GP should take a detailed history from Leanne and her parents and explore the 'trances' because they have experienced and witnessed them. This should determine whether an epileptic seizure is likely to have occurred. Diagnosis should not be based on the presence or absence of single features.

The GP should consider a history of absence seizures. Leanne is the right age and gender for this relatively common childhood epilepsy syndrome. The frequent occurrence of the daydreams and the fact that they interrupt her activities are suspicious features of childhood-onset absence epilepsy. The positive family history is supportive but not diagnostic of this epilepsy syndrome. Childhood-onset absence epilepsy is classified as an idiopathic (presumed genetic) generalised epilepsy.

The GP could potentially confirm the diagnosis in the surgery. Children with typical absence seizures will often experience one of their absences during hyperventilation (over-breathing). However, hyperventilation usually has to be performed well and for at least 3 minutes to induce an absence.

The GP should refer Leanne to a general paediatrician with an interest in epilepsy or a paediatric neurologist to establish the diagnosis.

Relevant recommendations

A detailed history should be taken from the child, young person or adult and an eyewitness to the attack, where possible, to determine whether or not an epileptic seizure is likely to have occurred. **[1.5.4]**

The clinical decision as to whether an epileptic seizure has occurred should then be based on the combination of the description of the attack and different symptoms. Diagnosis should not be based on the presence or absence of single features. **[1.5.5]**

The diagnosis of epilepsy in children and young people should be established by a specialist paediatrician with training and expertise in epilepsy. **[1.5.2]**

The paediatrician or paediatric neurologist takes a full history and examines Leanne. The history reveals that she is experiencing these episodes many times every day. When Leanne is encouraged to hyperventilate in the clinic, she has an absence during hyperventilation.

1.2 Question

What further investigations should the paediatrician or paediatric neurologist request, and what considerations should be taken into account?

1.2 Answer

First, information should be given to Leanne and her parents about the reasons for further tests, and they should be carried out in a child-centred environment.

An electroencephalogram (EEG) should be arranged and Leanne should have this test soon after it has been requested. Because the paediatrician or paediatric neurologist suspects that her seizures are epileptic in origin, the EEG should be performed to support a diagnosis of epilepsy. It should not be used in isolation to make a diagnosis of epilepsy. The healthcare professionals carrying out the EEG should encourage Leanne to hyperventilate, because this is one of the provocation techniques always undertaken during an EEG.

Relevant recommendations

Information should be provided to children, young people and adults and families and/or carers as appropriate on the reasons for tests, their results and meaning, the requirements of specific investigations, and the logistics of obtaining them. **[1.6.1]**

All investigations for children should be performed in a child-centred environment. **[1.6.2]**

Children, young people and adults requiring an EEG should have the test performed soon after it has been requested. **[1.6.3]**

An EEG should be performed only to support a diagnosis of epilepsy in adults in whom the clinical history suggests that the seizure is likely to be epileptic in origin. **[1.6.4]**

An EEG should be performed only to support a diagnosis of epilepsy in children and young people. If an EEG is considered necessary, it should be performed after the second epileptic seizure but may, in certain circumstances, as evaluated by the specialist, be considered after a first epileptic seizure. **[1.6.5]**

The EEG should not be used in isolation to make a diagnosis of epilepsy. **[1.6.8]**

Photic stimulation and hyperventilation should remain part of standard EEG assessment. The child, young person or adult and family and/or carer should be made aware that such activation procedures may induce a seizure and they have a right to refuse. **[1.6.18]**

The EEG is abnormal, as in the vast majority of children with childhood-onset absence epilepsy. It 'captures' an absence seizure, particularly during hyperventilation.

1.3 Question

The diagnosis of childhood-onset absence epilepsy is confirmed. What are the next steps?

1.3 Answer

The paediatrician or paediatric neurologist should discuss the diagnosis with Leanne and her parents. The discussion should focus on the likely cause (genetic), potential implications of frequent absences for Leanne (impaired concentration and therefore impaired learning, possible injury with continuing seizures), low risk of her developing other types of seizures (particularly tonic−clonic seizures) and likely response to anti-epileptic medication. This information should be provided in a suitable format. Details of the different sources of information available from voluntary organisations should also be given.

Adequate time should be set aside for the discussion. Leanne and her parents should know how to contact a named individual of the healthcare team when further information is needed. Information on sudden unexpected death in epilepsy (SUDEP) should be included within the literature given to Leanne's parents.

A comprehensive care plan should be agreed with Leanne and her parents. This should include lifestyle as well as medical issues.

Relevant recommendations

Children, young people and adults with epilepsy and their families and/or carers should be given, and have access to sources of, information about (where appropriate):

- epilepsy in general
- diagnosis and treatment options
- medication and side effects
- seizure type(s), triggers and seizure control
- management and self-care
- risk management
- first aid, safety and injury prevention at home and at school or work
- psychological issues
- social security benefits and social services
- insurance issues

- education and healthcare at school
- employment and independent living for adults
- importance of disclosing epilepsy at work, if relevant (if further information or clarification is needed, voluntary organisations should be contacted)
- road safety and driving
- prognosis
- sudden death in epilepsy (SUDEP)
- status epilepticus
- lifestyle, leisure and social issues (including recreational drugs, alcohol, sexual activity and sleep deprivation)
- family planning and pregnancy
- voluntary organisations, such as support groups and charitable organisations, and how to contact them. **[1.3.1]**

Information should be provided in formats, languages and ways that are suited to the child, young person or adult's requirements. Consideration should be given to developmental age, gender, culture and stage of life of the person. **[1.3.3]**

If children, young people and adults, and their families and/or carers, have not already found high-quality information from voluntary organisations and other sources, healthcare professionals should inform them of different sources (using the Internet, if appropriate: see, for example, the website of the Joint Epilepsy Council of the UK and Ireland, www.jointepilepsycouncil.org.uk). **[1.3.4]**

Adequate time should be set aside in the consultation to provide information, which should be revisited on subsequent consultations. **[1.3.5]**

The child, young person or adult with epilepsy and their family and/or carers as appropriate should know how to contact a named individual when information is needed. This named individual should be a member of the healthcare team and be responsible for ensuring that the information needs of the child, young person or adult and/or their family and/or carers are met. **[1.3.8]**

The possibility of having seizures should be discussed, and information on epilepsy should be provided before seizures occur, for children, young people and adults at high risk of developing seizures (such as after severe brain injury), with a learning disability, or who have a strong family history of epilepsy. **[1.3.9]**

Information on SUDEP should be included in literature on epilepsy to show why preventing seizures is important. Tailored information on the person's relative risk of SUDEP should be part of the counselling checklist for children, young people and adults with epilepsy and their families and/or carers. **[1.3.11]**

All children, young people and adults with epilepsy should have a comprehensive care plan that is agreed between the person, family and/or carers where appropriate, and primary care and secondary care providers. This should include lifestyle issues as well as medical issues. **[1.8.2]**

1.4 Question

What should the discussion around medication include, and what AEDs may be prescribed?

1.4 Answer

The discussion on anti-epileptic medication should include the different medications that are available, and specifically ethosuximide, sodium valproate and lamotrigine, the evidence base for using these medications and their common and potentially unwanted side effects. Discussion should also include the likely outcome or prognosis of the epilepsy and specifically that it will go into spontaneous remission (that is, it will 'go away'). The family should also be referred to a paediatric epilepsy nurse who can provide information and guidance on lifestyle and other non-medical issues.

Ethosuximide or sodium valproate should be offered as a first-line treatment. Be aware of the teratogenic risks of sodium valproate. Offer lamotrigine if the first-line AEDs are unsuitable, ineffective or not tolerated. If the first two AEDs are ineffective, consider a combination of two of these three AEDs as adjunctive treatment. If adjunctive treatment is ineffective or not tolerated, discuss with or refer to a tertiary epilepsy specialist and consider clobazam, clonazepam, levetiracetam, topiramate or zonisamide.

Do not offer carbamazepine, gabapentin, oxcarbazepine, phenytoin, pregabalin, tiagabine or vigabatrin.

Relevant recommendations

Epilepsy specialist nurses (ESNs) should be an integral part of the network of care of children, young people and adults with epilepsy. The key roles of the ESNs are to support both epilepsy specialists and generalists, to ensure access to community and multi-agency services and to provide information, training and support to the child, young person or adult, families, carers and, in the case of children, others involved in the child's education, welfare and well-being. **[1.8.3]**

Offer ethosuximide or sodium valproate as first-line treatment to children, young people and adults with absence seizures. If there is a high risk of GTC seizures, offer sodium valproate first, unless it is unsuitable. Be aware of teratogenic risks of sodium valproate (see recommendation 1.9.1.10 of the NICE guideline). **[1.9.5.1]**

Offer lamotrigine [13] if ethosuximide and sodium valproate are unsuitable, ineffective or not tolerated. **[1.9.5.2]**

If two first-line AEDs (see recommendations 1.9.5.1 and 1.9.5.2) are ineffective in children, young people and adults with absence seizures, consider a combination of two of these three AEDs as adjunctive treatment: ethosuximide, lamotrigine [13] or sodium valproate. Be aware of teratogenic risks of sodium valproate (see recommendation 1.9.1.10 of the NICE guideline). **[1.9.5.3]**

If adjunctive treatment (see recommendation 1.9.5.3 of the NICE guideline) is ineffective or not tolerated, discuss with, or refer to, a tertiary epilepsy specialist and consider clobazam [13], clonazepam, levetiracetam [13], topiramate [13] or zonisamide [13.] **[1.9.5.4]**

Do not offer carbamazepine, gabapentin, oxcarbazepine, phenytoin, pregabalin, tiagabine or vigabatrin. **[1.9.5.5]**

[13] At the time of publication (January 2012), this drug did not have UK marketing authorisation for this indication and/or population (see appendix E of the NICE guideline for details). Informed consent should be obtained and documented.

Supporting information

Some ‘facts’ about childhood absence epilepsy

- It comprises approximately 5−10% of all epilepsy in childhood.
- Age at onset: range (3−9 years) and peak (4−7 years).
- More common in girls: approximately 3:1 (possibly slightly higher).
- Family history of epilepsy: more often than not because this is a genetic epilepsy.
- Seizure type: all have typical absence seizures; the usual duration of each seizure is 5−12 seconds. Approximately 5−10% of children may also have tonic−clonic seizures.
- EEG: characteristic pattern with 3 Hz (cycles per second) generalised spike and slow-wave discharges – seen during an absence seizure; this activity may also be seen between absences.
- Other comorbid problems: children do not usually have any associated learning difficulties or behaviour problems – providing the child has been diagnosed (and treated) relatively quickly.
- Anti-epileptic medications of choice (based on current evidence): ethosuximide (if the child has only ever had absence seizures) or sodium valproate if the child has had tonic−clonic seizures and absence seizures. Lamotrigine is another option but is less effective than ethosuximide or sodium valproate in treating the absence seizures (Glauser TA, Cnaan A, Shinnar S et al. Ethosuximide, valproic acid, and lamotrigine in childhood absence epilepsy. New England Journal of Medicine 2010; 362: 790−9).
- Response to anti-epileptic medication: approximately 70% will become seizure-free on medication (ethosuximide or sodium valproate).
- Long-term outcome: approximately 70% of children will remit spontaneously. There is a low risk (< 10%) of the person developing tonic−clonic seizures in early adulthood even if the absence seizures have entered remission and the child is off anti-epileptic medication

(This information was obtained from a Guideline Development Group member.)

Case scenario 2: Christopher

Presentation

Christopher, a 13-month-old infant, presents to the accident and emergency department (A&E) of his local hospital with his first prolonged tonic−clonic afebrile seizure. He has experienced three febrile seizures in the past. The first two occurred at eight months of age due to a chest infection. They were prolonged, lasting over 15 minutes, and he required emergency medication and a three-day admission. The third febrile seizure was at nine months and associated with an upper respiratory tract infection. It only affected the left side of Christopher's body and lasted for over 30 minutes before it was stopped with intravenous lorazepam and intravenous phenytoin.

A magnetic resonance imaging (MRI) brain scan and cerebrospinal fluid analysis are normal. His early development has been normal but he only started crawling at 12 months of age and he has no recognisable words. There is no family history of epilepsy.

2.1 Question

What should the A&E staff looking after Christopher consider as a possible diagnosis, and what investigations should be carried out to confirm this? Also what referrals, if any, should be made?

2.1 Answer

Christopher's history is unusual for simple or complex febrile seizures and should raise the possibility of epilepsy. The features that suggest a diagnosis of Dravet syndrome (also known as severe myoclonic epilepsy of infancy) include age at onset, frequent and prolonged nature of the febrile seizures, recent development of a prolonged afebrile seizure and a background of possible developmental delay.

Christopher should be referred for an EEG and an opinion from a paediatric neurologist. The EEG may be normal or show non-specific abnormalities. The paediatric neurologist should consider the possibility of Dravet syndrome and undertake a blood test for DNA analysis to look for a mutation in the alpha (α) subunit of the first neuronal sodium channel (SCNIA) gene. A mutation is found in at least 80% of children (and adults) with Dravet syndrome.

An early diagnosis of Dravet syndrome establishes the cause of the Christopher's epilepsy and therefore avoids the need for him to undergo a number of investigations, some of which may be invasive.

Relevant recommendations

Administer intravenous lorazepam as first-line treatment in hospital in children, young people and adults with ongoing generalised tonic–clonic seizures (convulsive status epilepticus). Administer intravenous diazepam if intravenous lorazepam is unavailable, or buccal midazolam if unable to secure immediate intravenous access. Administer a maximum of two doses of the first-line treatment (including pre-hospital treatment). See also the suggested protocols in appendix F of the NICE guideline. **[1.14.2.2]**

If seizures continue, administer intravenous phenobarbital or phenytoin as second-line treatment in hospital in children, young people and adults with ongoing generalised tonic–clonic seizures (convulsive status epilepticus). See also the suggested protocols in appendix F of the NICE guideline. **[1.14.2.3]**

Neuroimaging should be used to identify structural abnormalities that cause certain epilepsies. **[1.6.19]**

MRI should be the imaging investigation of choice in children, young people and adults with epilepsy. **[1.6.20]**

MRI is particularly important in those:

- who develop epilepsy before the age of 2 years or in adulthood
- who have any suggestion of a focal onset on history, examination or EEG (unless clear evidence of benign focal epilepsy)
- in whom seizures continue in spite of first-line medication. **[1.6.21]**

The diagnosis of epilepsy in children and young people should be established by a specialist paediatrician with training and expertise in epilepsy. **[1.5.2]**

Discuss with, or refer to, a tertiary paediatric epilepsy specialist when a child presents with suspected Dravet syndrome. **[1.9.9.1]**

Supporting information

Febrile seizures are common, occurring in approximately 2−5% of children. Most occur between the first and third (but up to the fifth) year of life, are classified as simple, are generalised tonic−clonic in nature and last less than 10 minutes. Complex febrile seizures are less common, typically last longer than 15 minutes and may affect only one side of the body. Thirty per cent of children will experience a recurrence. Afebrile seizures (epilepsy) may subsequently develop in up to 5% of children who have experienced recurrent febrile seizures but this is usually many years later.

(This information was obtained from a Guideline Development Group member.)

2.2 Question

What else should be considered, and what information should be provided?

2.2 Answer

The diagnosis enables the paediatric neurologist to inform and counsel the family from an early stage about the medium- and long-term outcome of this genetic epilepsy and the likely (low) risk of other children being similarly affected. Children with Dravet syndrome usually experience multiple types of seizures, including focal, tonic−clonic, absence and myoclonic. The seizures are typically difficult to control and rarely if ever remit spontaneously. Affected children may also have moderate or severe learning and speech and language impairments. Giving this information helps parents to understand and accept their child's problems early, and to better plan their education and their own immediate and future family life.

The paediatric neurologist and other healthcare professionals caring for Christopher should adopt a consulting style that enables Christopher and his family to participate as partners in all decisions about his healthcare.

Relevant recommendations

Children, young people and adults with epilepsy and their families and/or carers should be empowered to manage their condition as well as possible. **[1.2.1]**

Healthcare professionals should adopt a consulting style that enables the child, young person or adult with epilepsy, and their family and/or carers as appropriate, to participate as partners in all decisions about their healthcare, and take fully into account their race, culture and any specific needs. **[1.1.1]**

2.3 Question

What AED medication should the paediatric neurologist prescribe for Christopher?

2.3 Answer

The paediatric neurologist should consider sodium valproate or topiramate as a first-line treatment. They should discuss the treatment with a tertiary epilepsy specialist if first-line treatments are ineffective or not tolerated, and consider clobazam or stiripentol as adjunctive treatment.

They should not offer carbamazepine, gabapentin, lamotrigine, oxcarbazepine, phenytoin, pregabalin, tiagabine or vigabatrin,

Relevant recommendations

Consider sodium valproate or topiramate [13] as first-line treatment in children with Dravet syndrome. **[1.9.9.2]**

Discuss with a tertiary epilepsy specialist if first-line treatments (see recommendation 1.9.9.2 of the NICE guideline) in children, young people and adults with Dravet syndrome are ineffective or not tolerated, and consider clobazam [13] or stiripentol as adjunctive treatment. **[1.9.9.3]**

Do not offer carbamazepine, gabapentin, lamotrigine, oxcarbazepine, phenytoin, pregabalin, tiagabine or vigabatrin. **[1.9.9.4]**

[13] At the time of publication (January 2012), this drug did not have UK marketing authorisation for this indication and/or population (see appendix E of the NICE guideline for details). Informed consent should be obtained and documented.

Supporting information

- Early diagnosis enables the paediatric neurologist to consider the early prescription of a specific anti-epileptic medication, stiripentol, which, in combination with one or two other anti-epileptic medications (sodium valproate or clobazam, or both), may be the best way to control Christopher's seizures. A ketogenic diet may also be a useful treatment option.
  (This information was obtained from a Guideline Development Group member.)

Case scenario 3: Charlie

Presentation

The parents of Charlie, an 8-month-old boy, visit the GP because they are concerned about some strange episodes he has been having over the previous three weeks. The episodes usually occur soon after he has woken but may sometimes happen as he is about to go to sleep. In these episodes his knees suddenly come up towards his chest, his arms bend at the elbows and his hands may clench. Each episode lasts a few seconds but they may occur repeatedly over many minutes. He is very upset after them.

Over the past two weeks Charlie has become more irritable and has stopped showing any interest in his toys. His birth and perinatal period were normal. There is no family history of epilepsy.

3.1 Question

What steps should Charlie's GP take next?

3.1 Answer

Charlie's GP should take a detailed history of precisely what happens in these episodes and ask if his parents have recorded any of them on a mobile phone. The GP should also include some questions about Charlie's development and examine his skin for any birthmarks. Charlie's GP should consider the possibility that he may be experiencing infantile spasms (also called 'salaam attacks'). If there is any likelihood of this diagnosis, he must refer Charlie urgently to the local paediatric department, either by telephone or by fax.

The GP should give information to Charlie's parents about how to recognise a seizure, first aid and the importance of reporting further attacks.

Relevant recommendations

It is recommended that all children and young people who have had a first nonfebrile seizure should be seen as soon as possible[7] by a specialist in the management of the epilepsies to ensure precise and early diagnosis and initiation of therapy as appropriate to their needs. **[1.4.6]**

Essential information on how to recognise a seizure, first aid, and the importance of reporting further attacks should be provided to a child, young person or adult who has experienced a possible first seizure, and their family/carer/parent as appropriate. This information should be provided while the child, young person or adult is awaiting a diagnosis and should also be provided to their family and/or carers. **[1.4.9]**

A detailed history should be taken from the child, young person or adult and an eyewitness to the attack, where possible, to determine whether or not an epileptic seizure is likely to have occurred. **[1.5.4]**

Discuss with, or refer to, a tertiary paediatric epilepsy specialist when an infant presents with infantile spasms. **[1.9.8.1]**

7 The Guideline Development Group considered that with a recent onset suspected seizure, referrals should be urgent, meaning that patients should be seen within 2 weeks..

There is a possibility that Charlie may have experienced infantile spasms. Charlie's GP refers him to his local paediatric department urgently.

3.2 Question

What should the paediatrician or paediatric neurologist do next?

3.2 Answer

The paediatrician or paediatric neurologist should see Charlie as soon as possible because he may have infantile spasms (West syndrome). Examination should include measurement of Charlie's head circumference, ultraviolet examination of his skin (to look for evidence of tuberous sclerosis) and a brief assessment of his development and visual behaviour.

The paediatrician is fairly sure that Charlie is experiencing infantile spasms and may have West syndrome.

Relevant recommendations

The diagnosis of epilepsy in children and young people should be established by a specialist paediatrician with training and expertise in epilepsy. **[1.5.2]**

The clinical decision as to whether an epileptic seizure has occurred should then be based on the combination of the description of the attack and different symptoms. Diagnosis should not be based on the presence or absence of single features. **[1.5.5]**

In a child, young person or adult presenting with an attack, a physical examination should be carried out. This should address their cardiac, neurological and mental status, and should include a developmental assessment where appropriate. **[1.4.8]**

3.3 Question

What further investigations should Charlie have, and what information should be given to his parents about these?

3.3 Answer

An EEG should be carried out as soon as possible and preferably within 24−48 hours. Ideally, it should be performed in both the waking and sleeping state. Charlie should also have an MRI brain scan. Depending on the results of the MRI, additional investigations may be necessary, including blood, urine and cerebrospinal fluid (CSF) analyses.

Charlie's parents should be given information on the reasons for these tests, their results and meaning. The requirements and logistics for each test should also be explained.

Relevant recommendations

Information should be provided to children, young people and adults and families and/or carers as appropriate on the reasons for tests, their results and meaning, the requirements of specific investigations, and the logistics of obtaining them. **[1.6.1]**

All investigations for children should be performed in a child-centred environment. **[1.6.2]**

Children, young people and adults requiring an EEG should have the test performed soon after it has been requested. **[1.6.3]**

An EEG should be performed only to support a diagnosis of epilepsy in children and young people. If an EEG is considered necessary, it should be performed after the second epileptic seizure but may, in certain circumstances, as evaluated by the specialist, be considered after a first epileptic seizure. **[1.6.5]**

The EEG should not be used in isolation to make a diagnosis of epilepsy. **[1.6.8]**

Repeated standard EEGs should not be used in preference to sleep or sleep-deprived EEGs. **[1.6.13]**

Neuroimaging should be used to identify structural abnormalities that cause certain epilepsies. **[1.6.19]**

MRI should be the imaging investigation of choice in children, young people and adults with epilepsy. **[1.6.20]**

MRI is particularly important in those:

- who develop epilepsy before the age of 2 years or in adulthood
- who have any suggestion of a focal onset on history, examination or EEG (unless clear evidence of benign focal epilepsy)
- in whom seizures continue in spite of first-line medication. **[1.6.21]**

In children and young people, other investigations, including blood and urine biochemistry, should be undertaken at the discretion of the specialist to exclude other diagnoses, and to determine an underlying cause of the epilepsy. **[1.6.28]**

Charlie's EEG shows hypsarrhythmia, the characteristic EEG patterns seen in infants with infantile spasms, thereby confirming that he has West syndrome. The long-term developmental outcome of infants with West syndrome is primarily dependent on the underlying cause. However, because West syndrome is a type of 'epileptic encephalopathy', which means that the frequency of the spasms and the EEG appearance of hypsarrhythmia may adversely affect the long-term developmental outcome, it is important to treat infants as soon as the diagnosis of infantile spasms (West syndrome) is made.

3.4 Question

Charlie's MRI brain scan will be undertaken in a few days. What treatment options should the paediatrician or paediatric neurologist offer his parents, and what information should they be given?

3.4 Answer

The paediatrician or paediatric neurologist should offer either vigabatrin or a corticosteroid.

Charlie's parents should be given information on epilepsy in general as well as on infantile spasms. Medications, side effects, management, education and SUDEP should also be discussed. Adequate time should be set aside at consultation to provide this information and it should be given in a format suited to Charlie's parents.

Relevant recommendations

Offer a steroid (prednisolone or tetracosactide [13]) or vigabatrin as first-line treatment to infants with infantile spasms that are not due to tuberous sclerosis. Carefully consider the risk–benefit ratio when using vigabatrin or steroids. **[1.9.8.2]**

Children, young people and adults with epilepsy and their families and/or carers should be given, and have access to sources of, information about (where appropriate):

- epilepsy in general
- diagnosis and treatment options
- medication and side effects
- seizure type(s), triggers and seizure control
- management and self-care
- risk management
- first aid, safety and injury prevention at home and at school or work
- psychological issues
- social security benefits and social services
- insurance issues
- education and healthcare at school
- employment and independent living for adults
- importance of disclosing epilepsy at work, if relevant (if further information

or clarification is needed, voluntary organisations should be contacted)

- road safety and driving
- prognosis
- sudden death in epilepsy (SUDEP)
- status epilepticus
- lifestyle, leisure and social issues (including recreational drugs, alcohol, sexual activity and sleep deprivation)
- family planning and pregnancy
- voluntary organisations, such as support groups and charitable organisations, and how to contact them. **[1.3.1]**

Information should be provided in formats, languages and ways that are suited to the child, young person or adult's requirements. Consideration should be given to developmental age, gender, culture and stage of life of the person. **[1.3.3]**

Adequate time should be set aside in the consultation to provide information, which should be revisited on subsequent consultations. **[1.3.5]**

13 At the time of publication (January 2012), this drug did not have UK marketing authorisation for this indication and/or population (see appendix E for details of the NICE guideline). Informed consent should be obtained and documented.

Supporting information

Additional information about West syndrome:

- Precise incidence and prevalence figures are difficult to determine; however, it is thought to affect approximately 1 in 1800−2500 children under 1 year of age; each year in Great Britain about 350−400 children under a year will develop West syndrome.
- Boys are affected slightly more often than girls (approximately 1.4:1).
- It develops between 1 and 13 months of age, but rarely less than 3 months or more than 11 months; the peak age at onset is 6−8 months.
- The characteristic seizure type is the 'epileptic spasm'. This typically occurs in clusters and most commonly after the infant has just woken from sleep (night- or day-time nap)
- The EEG shows a characteristic pattern – hypsarrhythmia, which may be 'classical' or 'modified'.
- There are more than 50 causes of West syndrome; the more common causes include tuberous sclerosis complex, as a sequel to neonatal hypoxic-ischaemic encephalopathy, cerebral dysplasia and chromosomal abnormalities (including Down syndrome).
- It may be misdiagnosed as colic.
- The developmental outcome is primarily determined by the cause of the syndrome. However, a delayed diagnosis and/or initiation of appropriate treatment of many weeks may contribute to a poorer outcome.

(This information was obtained from a Guideline Development Group member.)

Case scenario 4: Suzanne

Presentation

Suzanne, a 10-year-old girl presents to A&E reported to have had a generalised tonic−seizure while on an aeroplane from Australia. It has stopped by the time she is seen by the ambulance staff, and she has fully recovered by the time she reaches the hospital.

Past medical history

Suzanne's delivery was normal and full term. Early developmental milestones were achieved at a similar time to her older sister. She had recurrent otitis media between 2 and 5 years of age. There have been no concerns since starting school.

On examination

Suzanne is fully orientated in time and space. She is apyrexial. There is no focal neurological deficit.

4.1 Question

What should the casualty officer do?

4.1 Answer

Suzanne should be seen by the paediatrician on duty before discharge if full examination reveals no concern. Her parents should be given advice on what to do if a further seizure occurs. An appointment should be offered within 2 weeks with a designated paediatrician (with an expertise in epilepsy) for further evaluation and possible diagnosis.

Relevant recommendations

Children, young people and adults presenting to an Accident and Emergency department following a suspected seizure should be screened initially. This should be done by an adult or paediatric physician with onward referral to a specialist [6] when an epileptic seizure is suspected or there is diagnostic doubt. **[1.4.1]**

It is recommended that all children and young people who have had a first non-febrile seizure should be seen as soon as possible 7 by a specialist in the management of the epilepsies to ensure precise and early diagnosis and initiation of therapy as appropriate to their needs. **[1.4.6]**

Essential information on how to recognise a seizure, first aid, and the importance of reporting further attacks should be provided to a child, young person or adult who has experienced a possible [7] first seizure, and their family/carer/parent as appropriate. This information should be provided while the child, young person or adult is awaiting a diagnosis and should also be provided to their family and/or carers. **[1.4.9]**

6 For adults, a specialist is defined throughout as a medical practitioner with training and expertise in epilepsy. For children and young people, a specialist is defined throughout as a paediatrician with training and expertise in epilepsy.

7 The Guideline Development Group considered that with a recent onset suspected seizure, referrals should be urgent, meaning that patients should be seen within 2 weeks.

The paediatrician with an expertise in epilepsy obtains further history. Suzanne was sleeping when her mother noticed her face contorting to one side. Then her right arm stiffened and jerked, and soon this involved her whole body. It lasted for about 2 minutes, and she did not come around fully for a further 15 minutes.

Her mother reports no other history of note although when asked specifically about sleep disturbance, she mentions that Suzanne has come into her bedroom on two occasions in the previous 4 months being unable to speak or move her right arm. On both occasions this has appeared to resolve spontaneously within a couple of minutes.

There is no other previous history of note. Suzanne is progressing well at school and there is no family history.

4.2 Question

What investigation(s) should be performed?

4.2 Answer

A wake EEG should be requested. This shows spikes over the centrotemporal areas bilaterally. A diagnosis of benign epilepsy with centrotemporal spikes is made. The paediatrician explains there is no need for neuroimaging.

Relevant recommendations

Information should be provided to children, young people and adults and families and/or carers as appropriate on the reasons for tests, their results and meaning, the requirements of specific investigations, and the logistics of obtaining them. **[1.6.1]**

All investigations for children should be performed in a child-centred environment. **[1.6.2]**

Children, young people and adults requiring an EEG should have the test performed soon after it has been requested. **[1.6.3]**

An EEG should be performed only to support a diagnosis of epilepsy in children and young people. If an EEG is considered necessary, it should be performed after the second epileptic seizure but may, in certain circumstances, as evaluated by the specialist, be considered after a first epileptic seizure. **[1.6.5]**

The EEG should not be used in isolation to make a diagnosis of epilepsy. **[1.6.8]**

An EEG may be used to help determine seizure type and epilepsy syndrome in children, young people and adults in whom epilepsy is suspected. This enables them to be given the correct prognosis. **[1.6.9]**

When a standard EEG has not contributed to diagnosis or classification, a sleep EEG should be performed. **[1.6.14]**

An MRI is particularly recommended in those who have any suggestions of focal onset on history, examination or EEG (unless there is clear evidence of benign focal epilepsy) **[1.6.21]**

Supporting information

Benign epilepsy with centrotemporal spikes is probably the most common form of epilepsy in childhood. It has most common onset between 5 and 10 years. Typically children present with nocturnal seizures, either focal involving the mouth and face, or generalised. This epilepsy carries an excellent prognosis, with all children reported to enter remission by 14 years of age. Treatment may be thought to be unnecessary in view of the guaranteed remission. However, there is still the risk of recurrent generalised seizures, which carry the risk of morbidity, and 10−20% of children with this epilepsy experience seizures exclusively from the awake state. Therefore, the decision to treat will depend on the degree to which the seizures are interfering with the child's life. The treating physician should decide with the family and child whether treatment is desired. They should take into account that there is likely to be an excellent response to AEDs and that these will not be needed long term.

(This information was obtained from a Guideline Development Group member.)

Next steps for management

4.3 Question

What other information should be given to the family so that a decision about treatment can be made?

4.3 Answer

The diagnosis should be fully explained to Suzanne and her family. This includes the likely prognosis and the debate around whether or not treatment is required. The risks of epilepsy (including injury and SUDEP) versus the risks of treatment (side effects or idiosyncratic reactions) should be discussed. A decision about whether treatment is required will be based on frequency of seizures, risk and the wishes of Suzanne and her parents. If they decide that she should have treatment, then there should be a discussion as to whether the family wish to start treatment with carbamazepine or lamotrigine.

Relevant recommendations

Children, young people and adults with epilepsy and their families and/or carers should be given, and have access to sources of, information about (where appropriate):

- epilepsy in general
- diagnosis and treatment options
- medication and side effects
- seizure type(s), triggers and seizure control
- management and self-care
- risk management
- first aid, safety and injury prevention at home and at school or work
- psychological issues
- social security benefits and social services
- insurance issues
- education and healthcare at school
- employment and independent living for adults
- importance of disclosing epilepsy at work, if relevant (if further information or clarification is needed, voluntary organisations should be contacted)
- road safety and driving
- prognosis
- sudden death in epilepsy (SUDEP)
- status epilepticus
- lifestyle, leisure and social issues (including recreational drugs, alcohol,

sexual activity and sleep deprivation)

- family planning and pregnancy
- voluntary organisations, such as support groups and charitable organisations, and how to contact them. **[1.3.1]**

The time at which this information should be given will depend on the certainty of the diagnosis, and the need for confirmatory investigations. **[1.3.2]**

Adequate time should be set aside in the consultation to provide information, which should be revisited on subsequent consultations. **[1.3.5]**

Discuss with the child or young person, and their family and/or carers, whether AED treatment for benign epilepsy with centrotemporal spikes, Panayiotopoulos syndrome or late-onset childhood occipital epilepsy (Gastaut type) is indicated. **[1.9.11.1]**

Offer carbamazepine [13] or lamotrigine [13] as first-line treatment to children and young people with benign epilepsy with centrotemporal spikes, Panayiotopoulos syndrome or late-onset childhood occipital epilepsy (Gastaut type). **[1.9.11.2]**

13 At the time of publication (January 2012), this drug did not have UK marketing authorisation for this indication and/or population (see appendix E for details). Informed consent should be obtained and documented.

4.4 Question

How long should the treatment be continued for?

4.4 Answer

The prognosis for this syndrome is that children are likely to be in remission by the age of 14 years. Therefore, should Suzanne remain seizure-free, a discussion should be undertaken with the family about the best time for her medication to be gradually withdrawn and then stopped, probably around the age of 13 years.

Other implementation tools

NICE has developed tools to help organisations implement the clinical guideline on the epilepsies (listed below). These are available on the NICE website (www.nice.org.uk/guidance/CG137).

- Costing statement – details of the likely costs and savings when the cost impact of the guideline is not considered to be significant.
- Audit support – for monitoring local practice.
- Pharmacological treatment tables – tables from appendix E of the NICE guideline separated for ease of use and printing.
- Slide set - educational slide set which highlight the key recommendations.
- Online educational tool – developed in conjunction with BMJ Learning, the interactive module uses interactive case histories to improve user's knowledge of the guidance. The tools are free to use and open to all. You will need to provide your email address and a password to register with BMJ Learning.

A practical guide to implementation, 'How to put NICE guidance into practice: a guide to implementation for organisations', is also available (www.nice.org.uk/usingguidance/implementationtools).

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