The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care (partial update of NICE clinical guideline 20)

NICE guideline
Draft for consultation, January 2011

If you wish to comment on this version of the guideline, please be aware that all the supporting information and evidence is contained in the full version.
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This guidance is a partial update of NICE clinical guideline 20 (published October 2004) and will replace it. This guidance also updates NICE technology appraisal 76 (published March 2004) and NICE technology appraisal 79 (published April 2004) and will replace them. It was produced by the National Clinical Guideline Centre for Acute and Chronic Conditions.

New recommendations have been added for the pharmacological treatment of people with epilepsy, including the use of ketogenic diet.

Where recommendations are shaded in grey and end [2004] the evidence has not been updated since the original guideline. Yellow shading in these recommendations indicates where wording changes have been made for the purposes of clarification only. If the amendment has resulted in a change in meaning, the reason for the amendment is also provided using a footnote and the recommendation marked as [2004, amended 2011].

You are invited to comment on the new and updated recommendations in this guideline only. These are marked as [2011] if the evidence has been reviewed but no change has been made to the recommendation or [new 2011] if the evidence has been reviewed and the recommendation has been added or updated.

Appendix H contains recommendations from the 2004 guideline that NICE proposes deleting in the 2011 update. This is because the evidence has been reviewed and the recommendation has been updated or because NICE has updated other relevant guidance and has replaced the original recommendations. Where there are replacement recommendations based on a review of the evidence, this information is provided. Where there is no replacement recommendation, an explanation for the proposed deletion is given. You are invited to comment on the deleted recommendations as part of the consultation on the 2011 update.

The original NICE guideline and supporting documents are available from www.nice.org.uk/guidance/CG20
**Introduction**

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 260,000 and 416,000 people in England and Wales. 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).

Newer and more expensive AEDs are now being prescribed, and with an increase in treatment costs likely in coming years it is essential to ensure that AEDs with proven clinical and cost effectiveness are identified. The evidence used to develop NICE clinical guideline 20 and the existing NICE technology appraisal guidance on epilepsy (‘Newer drugs for epilepsy in adults’ [NICE technology appraisal guidance 76] and ‘Newer drugs for epilepsy in children’ [NICE technology appraisal guidance 79]) showed no difference in effectiveness between newer and older AEDs, or between the newer drugs (as monotherapy) for seizure control. However, a recent large multicentre trial (the SANAD trial) evaluating newer drugs in newly diagnosed epilepsy (accepting some limitations) suggested that sodium valproate should be the drug of choice in generalised and unclassifiable epilepsies, and lamotrigine in partial epilepsies. It was therefore considered necessary to review new evidence regarding AEDs within an update of NICE clinical guideline 20 (which was published in 2004).

The guideline will assume that prescribers will use a drug’s summary of product characteristics to inform decisions made with individual patients.
This guideline recommends some drugs for indications for which they do not have a UK marketing authorisation at the date of publication, if there is good evidence to support that use. Where recommendations have been made for the use of drugs outside their licensed indications ('off-label use'), these drugs are marked with a footnote in the recommendations.
Person-centred care

This guideline offers best practice advice on the care of children, young people and adults with epilepsy.

Treatment and care should take into account people’s needs and preferences. People with epilepsy should have the opportunity to make informed decisions about their care and treatment, in partnership with their healthcare professionals. If people do not have the capacity to make decisions, healthcare professionals should follow the Department of Health’s advice on consent (available from www.dh.gov.uk/consent) and the code of practice that accompanies the Mental Capacity Act (summary available from www.publicguardian.gov.uk). In Wales, healthcare professionals should follow advice on consent from the Welsh Assembly Government (available from www.wales.nhs.uk/consent).

If the person is under 16, healthcare professionals should follow the guidelines in ‘Seeking consent: working with children’ (available from www.dh.gov.uk/consent).

Good communication between healthcare professionals and people with epilepsy (and their family and carers) is essential. It should be supported by evidence-based written information tailored to the person’s needs. Treatment and care, and the information people are given about it, should be culturally appropriate. It should also be accessible to people with additional needs such as physical, sensory or learning disabilities, and to people who do not speak or read English.

If the person agrees, families and carers should have the opportunity to be involved in decisions about treatment and care.

Families and carers should also be given the information and support they need.

Care of young people in transition between paediatric and adult services should be planned and managed according to the best practice guidance.

Adult and paediatric healthcare teams should work jointly to provide assessment and services to young people with epilepsy. Diagnosis and management should be reviewed throughout the transition process, and there should be clarity about who is the lead clinician to ensure continuity of care.
Key priorities for implementation

The following recommendations have been identified as priorities for implementation.

Diagnosis

- **All children, young people and adults** with a recent onset suspected seizure should be seen urgently\(^1\) by a specialist\(^2\). This is to ensure precise and early diagnosis and initiation of therapy as appropriate to their needs. [2004]

Management

- 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. [2004]

- **All children, young people and adults** with epilepsy should have a comprehensive care plan that is agreed between the **person**, family and/or carers as appropriate, and primary and secondary care providers. [2004]

- The AED (anti-epileptic drug) treatment strategy should be individualised according to the seizure type, epilepsy syndrome, co-medication and co-morbidity, the **child’s, young person’s or adult’s** lifestyle, and the preferences of the **person**, their family and/or carers as appropriate. [2004]

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\(^1\) The Guideline Development Group considered that ‘urgently’ meant being seen within 2 weeks.

\(^2\) 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.
Prolonged or repeated seizures and convulsive status epilepticus

- Administer buccal midazolam* as first-line treatment in children, young people and adults with prolonged or repeated seizures. Administer rectal diazepam* if preferred or if buccal midazolam* is not available. If intravenous access is already established and resuscitation facilities are available, administer intravenous lorazepam. [new 2011]

- Only prescribe rectal diazepam* or buccal midazolam* for use in the community for children, young people and adults who have had a previous episode of prolonged or serial convulsive seizures. [new 2011]

Special considerations for women and girls of childbearing potential

- Women and girls with epilepsy and their partners, as appropriate, must be given accurate information and counselling about contraception, conception, pregnancy, caring for children, breastfeeding and menopause. [2004]

Review and referral

- All children, young people and adults with epilepsy should have a regular structured review. In children and young people, this review should be carried out at least yearly (but may be between 3 and 12 months by arrangement) by a specialist. In adults, this review should be carried out at least yearly by either a generalist or specialist, depending on how well the epilepsy is controlled and/or the presence of specific lifestyle issues. [2004]

- At the review, children, young people and adults should have access to: written and visual information; counselling services; information about voluntary organisations; epilepsy specialist nurses; timely and appropriate investigations; referral to tertiary services, including surgery if appropriate. [2004]

- If seizures are not controlled and/or there is diagnostic uncertainty or treatment failure, children, young people and adults should be referred to tertiary services soon³ for further assessment. [2004]

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* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.

³ The Guideline Development Group considered that 'soon' meant being seen within 4 weeks.
1 Guidance

The following guidance is based on the best available evidence. The full guideline ([hyperlink to be added for final publication]) gives details of the methods and the evidence used to develop the guidance.

In this guideline, adults are defined as aged 18 years and older and children as aged 28 days to 11 years. Young people are defined as aged 12 to 17 years. Older people are defined as aged older than 65 years. However, it is recognised that there is a variable age range (15–19 years) at which care is transferred between child and adult health services by local healthcare trusts and primary care organisations.

Please see appendix G for definitions of abbreviations and a glossary of terms used throughout this guideline.

1.1 Principle of decision making

1.1.1 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. [2004]

1.2 Coping with epilepsy

1.2.1 Children, young people and adults with epilepsy and their families and/or carers should be empowered to manage their condition as well as possible. [2004]

1.2.2 Adults should receive appropriate information and education about all aspects of epilepsy. This may be best achieved and maintained through structured self-management plans. [2004]

1.2.3 In children and young people, self-management of epilepsy may be best achieved through active child-centred training models and interventions. [2004]
1.2.4 Healthcare professionals should highlight the Expert Patients Programme (www.expertpatients.co.uk) to children, young people and adults with epilepsy who wish to manage their condition more effectively. [2004, amended 2011]

1.3 Information

1.3.1 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)

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4 This web address has changed since the recommendation was published in 2004 and has been updated.
• family planning and pregnancy
• voluntary organisations, such as support groups and charitable organisations, and how to contact them. [2004]

1.3.2 The time at which this information should be given will depend on the certainty of the diagnosis, and the need for confirmatory investigations. [2004]

1.3.3 Information should be provided in formats, languages and ways that are suited to the child’s, young person’s or adult’s requirements. Consideration should be given to developmental age, gender, culture and stage of life of the person. [2004]

1.3.4 If children, young people and adults, and 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). [2004]

1.3.5 Adequate time should be set aside in the consultation to provide information, which should be revisited on subsequent consultations. [2004]

1.3.6 Checklists should be used to remind children, young people and adults, and healthcare professionals, about information that should be discussed during consultations. [2004]

1.3.7 Everyone providing care or treatment for children, young people and adults with epilepsy should be able to provide essential information. [2004]

1.3.8 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. [2004]

1.3.9 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. [2004]

1.3.10 Children, young people and adults with epilepsy should be given appropriate information before they make important decisions (for example, regarding pregnancy or employment). [2004]

Sudden unexpected death in epilepsy (SUDEP)

1.3.11 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. [2004]

1.3.12 The risk of SUDEP can be minimised by:

- optimising seizure control
- being aware of the potential consequences of nocturnal seizures. [2004]

1.3.13 Tailored information and discussion between the child, young person or adult with epilepsy, their family and/or carers (as appropriate) and healthcare professionals should take account of the small but definite risk of SUDEP. [2004]

1.3.14 Where families and/or carers have been affected by SUDEP, healthcare professionals should contact families and/or carers to offer their condolences, invite them to discuss the death, and offer referral to bereavement counselling and a SUDEP support group. [2004]
1.4 Following a first seizure

1.4.1 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\(^5\) when an epileptic seizure is suspected or there is diagnostic doubt. [2004]

1.4.2 Protocols should be in place that ensure proper assessment in the emergency setting for children, young people and adults presenting with an epileptic seizure (suspected or confirmed). [2004]

1.4.3 The information that should be obtained from the adult and/or family or carer after a suspected seizure is contained in appendix D. [2004]

1.4.4 The information that should be obtained from the child or young person and/or parent or carer after a suspected seizure is contained in appendix D. [2004]

1.4.5 It is recommended that all adults having a first seizure should be seen as soon as possible\(^6\) by a specialist in the management of the epilepsies to ensure precise and early diagnosis and initiation of therapy as appropriate to their needs. [2004]

1.4.6 It is recommended that all children and young people who have had a first non-febrile seizure should be seen as soon as possible\(^6\) by a specialist in the management of the epilepsies to ensure precise and early diagnosis and initiation of therapy as appropriate to their needs. [2004]

1.4.7 At the initial assessment for a recent onset seizure, the specialist should have access to appropriate investigations. [2004]

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\(^5\) 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.

\(^6\) 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.
1.4.8 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. [2004]

1.4.9 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. [2004]

1.5 Diagnosis

1.5.1 The diagnosis of epilepsy in adults should be established by a specialist medical practitioner with training and expertise in epilepsy. [2004]

1.5.2 The diagnosis of epilepsy in children and young people should be established by a specialist paediatrician with training and expertise in epilepsy. [2004]

1.5.3 Children, young people and adults and their families and/or carers should be given an opportunity to discuss the diagnosis with an appropriate healthcare professional. [2004]

1.5.4 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. [2004]

1.5.5 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. [2004]
1.5.6 It may not be possible to make a definite diagnosis of epilepsy. If the diagnosis cannot be clearly established, further investigations (see section 1.6) and/or referral to a tertiary centre (see recommendation 1.10.2) should be considered. Follow-up should always be arranged. [2004]

1.5.7 Where non-epileptic attack disorder is suspected, suitable referral should be made to psychological or psychiatric services for further investigation and treatment. [2004]

1.5.8 Prospective recording of events, including video recording and written descriptions, can be very helpful in reaching a diagnosis. [2004]

1.6 Investigations

1.6.1 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. [2004]

1.6.2 All investigations for children should be performed in a child-centred environment. [2004]

Electroencephalogram (EEG)

1.6.3 Children, young people and adults requiring an EEG should have the test performed soon after it has been requested. [2004]

1.6.4 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. [2004]

1.6.5 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

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seizure but may, in certain circumstances, as evaluated by the specialist, be considered after a first epileptic seizure. [2004]

1.6.6 An EEG should not be performed in the case of probable syncope because of the possibility of a false-positive result. [2004]

1.6.7 The EEG should not be used to exclude a diagnosis of epilepsy in a child, young person or adult in whom the clinical presentation supports a diagnosis of a non-epileptic event. [2004]

1.6.8 The EEG should not be used in isolation to make a diagnosis of epilepsy. [2004]

1.6.9 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. [2004]

1.6.10 In children, young people and adults presenting with a first unprovoked seizure, unequivocal epileptiform activity shown on EEG can be used to assess the risk of seizure recurrence. [2004]

1.6.11 For children, young people and adults in whom epilepsy is suspected, but who present diagnostic difficulties, specialist investigations should be available. [2004]

1.6.12 Repeated standard EEGs may be helpful when the diagnosis of the epilepsy or the syndrome is unclear. However, if the diagnosis has been established, repeat EEGs are not likely to be helpful. [2004]

1.6.13 Repeated standard EEGs should not be used in preference to sleep or sleep-deprived EEGs. [2004]

1.6.14 When a standard EEG has not contributed to diagnosis or classification, a sleep EEG should be performed. [2004]
1.6.15 In children and young people, a sleep EEG is best achieved through sleep deprivation or the use of melatonin. [2004, amended 2011]

1.6.16 Long-term video or ambulatory EEG may be used in the assessment of children, young people and adults who present diagnostic difficulties after clinical assessment and standard EEG. [2004]

1.6.17 Provocation by suggestion may be used in the evaluation of non-epileptic attack disorder. However, it has a limited role and may lead to false-positive results in some people. [2004]

1.6.18 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. [2004]

**Neuroimaging**

1.6.19 Neuroimaging should be used to identify structural abnormalities that cause certain epilepsies. [2004]

1.6.20 MRI should be the imaging investigation of choice in children, young people and adults with epilepsy. [2004]

1.6.21 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. [2004]

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8 The licence for use of melatonin in the UK has changed since the recommendation was published in 2004; therefore, the recommendation has been updated accordingly within this update, and the footnote that contained this information has been deleted.
1.6.22 Children, young people and adults requiring MRI should have the test performed soon. [2004]

1.6.23 Neuroimaging should not be routinely requested when a diagnosis of idiopathic generalised epilepsy has been made. [2004]

1.6.24 CT should be used to identify underlying gross pathology if MRI is not available or is contraindicated, and for children or young people in whom a general anaesthetic or sedation would be required for MRI but not CT. [2004]

1.6.25 In an acute situation, CT may be used to determine whether a seizure has been caused by an acute neurological lesion or illness. [2004]

Other tests

1.6.26 Measurement of serum prolactin is not recommended for the diagnosis of epilepsy. [2004]

1.6.27 In adults, appropriate blood tests (for example, plasma electrolytes, glucose, calcium) to identify potential causes and/or to identify any significant co-morbidity should be considered. [2004]

1.6.28 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. [2004]

1.6.29 A 12-lead ECG should be performed in adults with suspected epilepsy. [2004]

1.6.30 In children and young people, a 12-lead ECG should be considered in cases of diagnostic uncertainty. [2004]

1.6.31 In cases of diagnostic uncertainty, a referral to a cardiologist should be considered. [2004]

9 The Guideline Development Group considered that ‘soon’ meant being seen within 4 weeks.
### Neuropsychological assessment

1.6.32 Neuropsychological assessment should be considered in **children, young people and adults** in whom it is important to evaluate learning disabilities and cognitive dysfunction, particularly in regard to language and memory. [2004]

1.6.33 Referral for a neuropsychological assessment is indicated:

- when a **child, young person or adult** with epilepsy is having educational or occupational difficulties
- when an MRI has identified abnormalities in cognitively important brain regions
- when a **child, young person or adult** complains of memory or other cognitive deficits and/or cognitive decline. [2004]

### Classification

1.7.1 Epileptic seizures and epilepsy syndromes in **children, young people and adults** should be classified using a multi-axial diagnostic scheme. The axes that should be considered are: description of seizure (ictal phenomenology); seizure type; syndrome and aetiology. [2004]

1.7.2 The seizure type(s) and epilepsy syndrome, aetiology, and co-morbidity should be determined, because failure to classify the epilepsy syndrome correctly can lead to inappropriate treatment and persistence of seizures. [2004]

1.7.3 **Children, young people and adults** with epilepsy should be given information about their seizure type(s) and epilepsy syndrome, and the likely prognosis. [2004]
1.8  Management

1.8.1  Children, young people and adults with epilepsy should have an accessible point of contact with specialist services. [2004]

1.8.2  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. [2004]

1.8.3  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. [2004]

1.8.4  Healthcare professionals have a responsibility to educate others about epilepsy so as to reduce the stigma associated with it. They should provide information about epilepsy to all people who come into contact with children, young people and adults with epilepsy, including school staff, social care professionals and others. [2004]

1.9  Pharmacological treatment

Note: see appendix E for further details of pharmacological treatment.

1.9.1  General information about pharmacological treatment

1.9.1.1  Information that is provided about anti-epileptic drugs (AEDs) needs to be in the context of that provided by the manufacturer, for example, indications, side effects and licence status. [2004]

1.9.1.2  The AED treatment strategy should be individualised according to the seizure type, epilepsy syndrome, co-medication and co-
morbidity, the child’s, young person’s or adult’s lifestyle, and the preferences of the person and their family and/or carers as appropriate (see appendix E). [2004]

1.9.1.3 The diagnosis of epilepsy needs to be critically evaluated if events continue despite an optimal dose of a first-line AED. [2004]

1.9.1.4 Changing the formulation or brand of AED is not recommended because different preparations may vary in bioavailability or have different pharmacokinetic profiles and, thus, increased potential for reduced effect or excessive side effects. [2004]

1.9.1.5 Follow guidance in the summary of product characteristics (SPC) and ‘British national formulary’ (BNF; available at http://bnf.org) on the bioavailability and pharmacokinetic profiles of AEDs. [new 2011]

1.9.1.6 It is recommended that children, young people and adults should be treated with a single AED (monotherapy) wherever possible. If the initial treatment is unsuccessful, then monotherapy using another drug can be tried. Caution is needed during the changeover period. [2004]

1.9.1.7 If an AED has failed because of adverse effects or continued seizures, a second drug should be started (which may be an alternative first-line or second-line drug) and built up to an adequate or maximum tolerated dose and then the first drug should be tapered off slowly. [2004]

1.9.1.8 If the second drug is unhelpful, either the first or second drug may be tapered, depending on relative efficacy, side effects and how well the drugs are tolerated before starting another drug. [2004]

1.9.1.9 It is recommended that combination therapy (adjunctive or ‘add-on’ therapy) should only be considered when attempts at monotherapy with AEDs have not resulted in seizure freedom. If trials of
combination therapy do not bring about worthwhile benefits, treatment should revert to the regimen (monotherapy or combination therapy) that has proved most acceptable to the child, young person or adult, in terms of providing the best balance between effectiveness in reducing seizure frequency and tolerability of side effects. [2004]

1.9.1.10 If using carbamazepine, offer controlled-release carbamazepine preparations. [new 2011]

1.9.1.11 When prescribing sodium valproate to women and girls of present and future childbearing potential discuss the possible risk to an unborn child of malformation and neurodevelopmental delay particularly with high doses of this AED. [new 2011]

1.9.2 Initiation of pharmacological treatment

1.9.2.1 AED therapy should only be started once the diagnosis of epilepsy is confirmed, except in exceptional circumstances that require discussion and agreement between the prescriber, the specialist and the child, young person or adult and their family and/or carers as appropriate. [2004]

1.9.2.2 AED therapy should be initiated in adults on the recommendation of a specialist. [2004]

1.9.2.3 AED therapy in children and young people should be initiated by a specialist. [2004]

1.9.2.4 The decision to initiate AED therapy should be taken between the child, young person or adult, their family and/or carers (as appropriate) and the specialist after a full discussion of the risks and benefits of treatment. This discussion should take into account details of the person’s epilepsy syndrome, prognosis and lifestyle. [2004]
1.9.2.5 Treatment with AED therapy is generally recommended after a second epileptic seizure. [2004]

1.9.2.6 When possible, choose which AED to offer on the basis of the presenting epilepsy syndrome. If the epilepsy syndrome is not clear at presentation, base the decision on the presenting seizure type(s). [new 2011]

1.9.2.7 AED therapy should be considered and discussed with children, young people and adults and their family and/or carers as appropriate after a first unprovoked seizure if:

- the child, young person or adult has a neurological deficit
- the EEG shows unequivocal epileptic activity
- the child, young person or adult and/or their family and/or carers consider the risk of having a further seizure unacceptable
- brain imaging shows a structural abnormality. [2004]

1.9.2.8 It should be recognised that some children, young people and adults (through their families and/or carers, in some instances) may choose not to take AED therapy following a full discussion of the risks and benefits. [2004]

### 1.9.3 Pharmacological treatment of focal seizures

#### First-line treatment in children, young people and adults with newly diagnosed focal seizures

1.9.3.1 Offer carbamazepine, lamotrigine, oxcarbazepine or sodium valproate as first-line treatment to children, young people and adults with newly diagnosed focal seizures, unless they are unsuitable. If the first AED is ineffective or not tolerated, offer an alternative from these four AEDs. If the second well-tolerated AED is ineffective, consider adjunctive treatment. [new 2011]

1.9.3.2 Levetiracetam is not cost effective at current 2010 unit costs (estimated cost of a 1500 mg daily dose is £2.74). Therefore, offer levetiracetam if:
• cost-effective first-line treatments (carbamazepine, lamotrigine, oxcarbazepine or sodium valproate) are unsuitable, or
• acquisition costs are reduced by at least 50% as an alternative first-line treatment. [new 2011]

Adjunctive treatment in children, young people and adults with refractory focal seizures

1.9.3.3 Offer carbamazepine, clobazam*, gabapentin*, lamotrigine, levetiracetam, oxcarbazepine, sodium valproate or topiramate as adjunctive treatment to children, young people and adults with focal seizures if first-line treatments (see recommendations 1.9.3.1 and 1.9.3.2) are ineffective or not tolerated. [new 2011]

1.9.3.4 If adjunctive treatment (see recommendation 1.9.3.3) is ineffective or not tolerated, discuss with, or offer referral to, a tertiary epilepsy specialist. Other AEDs that may be considered are eslicarbazepine acetate*, lacosamide, phenobarbital, phenytoin, pregabalin*, tiagabine, vigabatrin and zonisamide*. Carefully consider the risk–benefit ratio when using vigabatrin because of the risk of an irreversible effect on visual fields. [new 2011]

1.9.4 Pharmacological treatment of newly diagnosed generalised tonic–clonic (GTC) seizures

First-line treatment in children, young people and adults with newly diagnosed GTC seizures

1.9.4.1 Offer lamotrigine or sodium valproate as first-line treatment to children, young people and adults with newly diagnosed GTC seizures. If they have myoclonic seizures, or are suspected of having juvenile myoclonic epilepsy (JME), offer sodium valproate first, unless it is unsuitable (see recommendation 1.9.13.1). [new 2011]

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
1.9.4.2 Consider carbamazepine and oxcarbazepine* but be aware of the risk of exacerbating myoclonic and/or absence seizures. [new 2011]

Adjunctive treatment in children, young people and adults with GTC seizures

1.9.4.3 Offer clobazam*, lamotrigine, levetiracetam, sodium valproate or topiramate as adjunctive treatment to children, young people and adults with GTC seizures if first-line treatments (see recommendations 1.9.4.1 and 1.9.4.2) are ineffective, not tolerated or unsuitable. [new 2011]

1.9.4.4 If there are absence or myoclonic seizures, or if JME is suspected, do not offer carbamazepine, gabapentin, oxcarbazepine, phenytoin, pregabalin, tiagabine or vigabatrin. [new 2011]

1.9.5 Pharmacological treatment of absence seizures

First-line treatment in children, young people and adults with absence seizures

1.9.5.1 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. [new 2011]

1.9.5.2 Offer lamotrigine* if ethosuximide and sodium valproate are ineffective, not tolerated or unsuitable. [new 2011]

Adjunctive treatment in children, young people and adults with absence seizures

1.9.5.3 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* or sodium valproate. [new 2011]

* At the time of publication (month year), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
1.9.5.4 If adjunctive treatment (see recommendation 1.9.5.3) is ineffective or not tolerated, discuss with, or refer to, a tertiary epilepsy specialist and consider clobazam*, clonazepam, levetiracetam*, topiramate* or zonisamide*. [new 2011]

1.9.5.5 Do not offer carbamazepine, gabapentin, oxcartazepine, phenytoin, pregabalin, tiagabine or vigabatrin. [new 2011]

1.9.6 Pharmacological treatment of myoclonic seizures

First-line treatment in children, young people and adults with myoclonic seizures

1.9.6.1 Offer sodium valproate as first-line treatment to children, young people and adults with newly diagnosed myoclonic seizures, unless it is unsuitable. [new 2011]

1.9.6.2 Consider levetiracetam* or topiramate* if sodium valproate is unsuitable or not tolerated. Be aware that topiramate* has a less favourable side-effect profile than levetiracetam* and sodium valproate. [new 2011]

Adjunctive treatment in children, young people and adults with myoclonic seizures

1.9.6.3 Offer levetiracetam, sodium valproate or topiramate* as adjunctive treatment to children, young people and adults with myoclonic seizures if first-line treatments (see recommendations 1.9.6.1 and 1.9.6.2) are ineffective or not tolerated. [new 2011]

1.9.6.4 If adjunctive treatment (see recommendation 1.9.6.3) is ineffective or not tolerated, discuss with, or refer to, a tertiary epilepsy specialist and consider clobazam*, clonazepam, piracetam or zonisamide*. [new 2011]

1.9.6.5 Do not offer carbamazepine, gabapentin, oxcartazepine, phenytoin, pregabalin, tiagabine or vigabatrin. [new 2011]

* At the time of publication (month year), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
1.9.7 Pharmacological treatment of tonic or atonic seizures

First-line treatment in children, young people and adults with tonic or atonic seizures

1.9.7.1 Offer sodium valproate as first-line treatment to children, young people and adults with tonic or atonic seizures. [new 2011]

Adjunctive treatment in children, young people and adults with tonic or atonic seizures

1.9.7.2 Offer lamotrigine* as adjunctive treatment to children, young people and adults with tonic or atonic seizures if first-line treatment with sodium valproate is ineffective or not tolerated. [new 2011]

1.9.7.3 Discuss with a tertiary epilepsy specialist if adjunctive treatment (see recommendation 1.9.7.2) is ineffective or not tolerated. Other AEDs that may be considered are rufinamide and topiramate*.

[new 2011]

1.9.7.4 Do not offer carbamazepine, gabapentin, oxcarbazepine, pregabalin, tiagabine or vigabatrin. [new 2011]

1.9.8 Pharmacological treatment of infantile spasms

First-line treatment in infants with infantile spasms

1.9.8.1 Discuss with, or refer to, a tertiary paediatric epilepsy specialist when an infant presents with infantile spasms. [new 2011]

1.9.8.2 Offer a steroid (prednisolone or tetracosactide*) or vigabatrin as first-line treatment to infants with infantile spasms that are not due to tuberous sclerosis. [new 2011]

1.9.8.3 Offer vigabatrin as first-line treatment to infants with infantile spasms due to tuberous sclerosis. If vigabatrin is ineffective, offer a steroid (prednisolone or tetracosactide*). [new 2011]

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
1.9.9  Pharmacological treatment of Dravet syndrome

First-line treatment in children and young people with Dravet syndrome
1.9.9.1  Consider sodium valproate or topiramate* as first-line treatment in children and young people with Dravet syndrome. [new 2011]

Adjunctive treatment in children and young people with Dravet syndrome
1.9.9.2  Refer children and young people with Dravet syndrome, in whom first-line treatment (see recommendation 1.9.9.1) has proved ineffective or not tolerated, to a tertiary paediatric epilepsy specialist for consideration of clobazam* or stiripentol as adjunctive treatment. [new 2011]

1.9.9.3  Do not offer carbamazepine, gabapentin, lamotrigine, oxcarbazepine, phenytoin, pregabalin, tiagabine or vigabatrin. [new 2011]

1.9.10  Pharmacological treatment of Lennox–Gastaut syndrome

First-line treatment in children and young people with Lennox–Gastaut syndrome
1.9.10.1  Offer sodium valproate as first-line treatment to children and young people with Lennox–Gastaut syndrome. [new 2011]

Adjunctive treatment in children, young people and adults with Lennox–Gastaut syndrome
1.9.10.2  Offer lamotrigine as adjunctive treatment to children, young people and adults with Lennox–Gastaut syndrome if first-line treatment with sodium valproate is ineffective or not tolerated. [new 2011]

1.9.10.3  Discuss with a tertiary epilepsy specialist if adjunctive treatment (see recommendation 1.9.10.2) is ineffective or not tolerated. Other AEDs that may be considered are rufinamide and topiramate. [new 2011]

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
1.9.10.4 Do not offer carbamazepine, gabapentin, oxcarbazepine, pregabalin, tiagabine or vigabatrin. [new 2011]

1.9.10.5 Only offer felbamate* in centres providing tertiary epilepsy specialist care and when treatment with all of the AEDs listed in recommendations 1.9.10.2 and 1.9.10.3 have proved ineffective or are not tolerated. [new 2011]

1.9.11 Pharmacological treatment of benign epilepsy with centrotemporal spikes, Panayiotopoulos syndrome or late-onset childhood occipital epilepsy (Gastaut type)

First-line treatment in children and young people with benign epilepsy with centrotemporal spikes, Panayiotopoulos syndrome or late-onset childhood occipital epilepsy (Gastaut type)

1.9.11.1 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. [new 2011]

1.9.11.2 Offer carbamazepine*, lamotrigine*, oxcarbazepine* or sodium valproate 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) when treatment is indicated. [new 2011]

1.9.11.3 Levetiracetam* is not cost effective at current 2010 unit costs (estimated cost of a 1500 mg daily dose is £2.74). Therefore, offer levetiracetam if:

- cost-effective first-line treatments (carbamazepine, lamotrigine, oxcarbazepine or sodium valproate) are unsuitable, or
- acquisition costs are reduced by at least 50% as an alternative first-line treatment. [new 2011]

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
1.9.12 Pharmacological treatment of idiopathic generalised epilepsy (IGE)

First-line treatment in children, young people and adults with IGE

1.9.12.1 Offer sodium valproate as first-line treatment to children, young people and adults with newly diagnosed IGE, particularly if there is a photoparoxysmal response on EEG. Offer lamotrigine* if sodium valproate is unsuitable or not tolerated. Be aware that lamotrigine can exacerbate myoclonic seizures. If JME is suspected see recommendation 1.9.13.1. [new 2011]

1.9.12.2 Consider topiramate*, but be aware that it has a less favourable side-effect profile than sodium valproate and lamotrigine*.

[new 2011]

Adjunctive treatment in children, young people and adults with IGE

1.9.12.3 Offer lamotrigine*, levetiracetam*, sodium valproate or topiramate* as adjunctive treatment to children, young people and adults with IGE if first-line treatments (see recommendations 1.9.12.1, 1.9.12.2) are ineffective or not tolerated. [new 2011]

1.9.12.4 If adjunctive treatment (see recommendation 1.9.12.3) is ineffective or not tolerated, discuss with, or refer to, a tertiary epilepsy specialist and consider clobazam*, clonazepam or zonisamide*.

[new 2011]

1.9.12.5 Do not offer carbamazepine, gabapentin, oxcarbazepine, phenytoin, pregabalin, tiagabine or vigabatrin. [new 2011]

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
1.9.13 Pharmacological treatment of juvenile myoclonic epilepsy (JME)

First-line treatment in children, young people and adults with JME

1.9.13.1 Offer sodium valproate as first-line treatment to children, young people and adults with newly diagnosed JME, unless it is unsuitable. [new 2011]

1.9.13.2 Consider lamotrigine*, levetiracetam* or topiramate* if sodium valproate is unsuitable or not tolerated. Be aware that topiramate has a less favourable side-effect profile than lamotrigine, levetiracetam and sodium valproate, and that lamotrigine can exacerbate myoclonic seizures. [new 2011]

Adjunctive treatment in children, young people and adults with JME

1.9.13.3 Offer lamotrigine*, levetiracetam, sodium valproate or topiramate* as adjunctive treatment to children, young people and adults with JME if first-line treatments (see recommendations 1.9.13.1, 1.9.13.2) are ineffective or not tolerated. [new 2011]

1.9.13.4 If adjunctive treatment (see recommendation 1.9.13.3) is ineffective or not tolerated, discuss with, or refer to, a tertiary epilepsy specialist and consider clobazam*, clonazepam or zonisamide*. [new 2011]

1.9.14 Pharmacological treatment of epilepsy with generalised tonic–clonic (GTC) seizures only

First-line treatment in children, young people and adults with epilepsy with GTC seizures only

1.9.14.1 Offer lamotrigine or sodium valproate as first-line treatment to children, young people and adults with epilepsy with GTC seizures

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
only. If they have myoclonic seizures, or are suspected of having JME, offer sodium valproate first, unless it is unsuitable (see recommendation 1.9.13.1). [new 2011]

1.9.14.2 Consider carbamazepine and oxcarbazepine* but be aware of the risk of exacerbating myoclonic and/or absence seizures. [new 2011]

Adjunctive treatment in children, young people and adults with GTC seizures only

1.9.14.3 Offer clobazam*, lamotrigine, levetiracetam, sodium valproate or topiramate as adjunctive treatment to children, young people and adults with epilepsy with GTC seizures only, if first-line treatments (see recommendation 1.9.14.1, 1.9.14.2) are ineffective, not tolerated or unsuitable. [new 2011]

1.9.15 Pharmacological treatment of childhood absence epilepsy, juvenile absence epilepsy or other absence epilepsy syndromes

First-line treatment in children, young people and adults with childhood absence epilepsy, juvenile absence epilepsy or other absence epilepsy syndromes

1.9.15.1 Offer ethosuximide or sodium valproate as first-line treatment to children, young people and adults with absence syndromes. If there is a high risk of GTC seizures, offer sodium valproate first. [new 2011]

1.9.15.2 Offer lamotrigine* if ethosuximide and sodium valproate are ineffective, not tolerated or unsuitable. [new 2011]

Adjunctive treatment in children, young people and adults with childhood absence epilepsy, juvenile absence epilepsy or other absence epilepsy syndromes

1.9.15.3 If two first-line AEDs (see recommendations 1.9.15.1 and 1.9.15.2) are ineffective, consider a combination of two of these three AEDs

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
as adjunctive treatment: ethosuximide, lamotrigine* or sodium valproate. [new 2011]

1.9.15.4 If adjunctive treatment (see recommendation 1.9.15.3) is ineffective or not tolerated, discuss with, or refer to, a tertiary epilepsy specialist and consider clobazam*, clonazepam, levetiracetam*, topiramate* or zonisamide*. [new 2011]

1.9.15.5 Do not offer carbamazepine, gabapentin, oxcarbazepine, phenytoin, pregabalin, tiagabine or vigabatrin. [new 2011]

1.9.16 Other epilepsy syndromes

1.9.16.1 Refer to a tertiary paediatric epilepsy specialist all children and young people with continuous spike and wave during slow sleep (CSWS), Landau–Kleffner syndrome (LKS) or myoclonic-astatic epilepsy (MAE). [new 2011]

1.9.17 Continuation of pharmacological treatment

1.9.17.1 Continuing AED therapy should be planned by the specialist. It should be part of the child’s, young person’s or adult’s agreed treatment plan, which should include details of how specific drug choices were made, drug dosage, possible side effects, and action to take if seizures persist. [2004]

1.9.17.2 The needs of the child, young person or adult and their family and/or carers as appropriate should be taken into account when healthcare professionals take on the responsibility of continuing prescribing. [2004]

1.9.17.3 If management is straightforward, continuing AED therapy can be prescribed in primary care if local circumstances and/or licensing allow. [2004]

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
1.9.17.4 The prescriber must ensure that the child, young person or adult and their family and/or carers as appropriate are fully informed about treatment including action to be taken after a missed dose or after a gastrointestinal upset. [2004]

1.9.17.5 Adherence to treatment can be optimised with the following:

- educating children, young people and adults and their families and/or carers in the understanding of their condition and the rationale of treatment
- reducing the stigma associated with the condition (see also section 1.2)
- using simple medication regimens
- positive relationships between healthcare professionals, the child, young person or adult with epilepsy and their family and/or carers. [2004]

1.9.17.6 Regular blood test monitoring in adults is not recommended as routine, and should be done only if clinically indicated. [2004]

1.9.17.7 Regular blood test monitoring in children and young people is not recommended as routine, and should be done only if clinically indicated and recommended by the specialist. [2004]

1.9.17.8 Indications for monitoring of AED blood levels are:

- detection of non-adherence to the prescribed medication
- suspected toxicity
- adjustment of phenytoin dose
- management of pharmacokinetic interactions
- specific clinical conditions, for example, status epilepticus, organ failure and pregnancy (see recommendation 1.15.3.9). [2011]
1.9.17.9 Examples of blood tests for adults include:

- before surgery – clotting studies in those on sodium valproate\(^{10}\)
- full blood count, electrolytes, liver enzymes, vitamin D levels, and other tests of bone metabolism (for example, serum calcium and alkaline phosphatase) every 2–5 years for adults taking enzyme-inducing drugs. [2004, amended 2011]

1.9.17.10 Asymptomatic minor abnormalities in test results are not necessarily an indication for changes in medication. [2004]

1.9.18 Withdrawal of pharmacological treatment

1.9.18.1 The decision to continue or withdraw medication should be taken by the child, young person or adult, their family and/or carers as appropriate, and the specialist after a full discussion of the risks and benefits of withdrawal. At the end of the discussion children, young people and adults, and their family and/or carers as appropriate, should understand their risk of seizure recurrence on and off treatment. This discussion should take into account details of the child’s, young person’s or adult’s epilepsy syndrome, prognosis and lifestyle. [2004]

1.9.18.2 Withdrawal of AEDs must be managed by, or be under the guidance of, the specialist. [2004]

1.9.18.3 The risks and benefits of continuing or withdrawing AED therapy should be discussed with children, young people and adults, and their families and/or carers as appropriate, who have been seizure free for at least 2 years (see appendix H\(^{11}\) of the full guideline). [2004]

1.9.18.4 When AED treatment is being discontinued in a child, young person or adult who has been seizure free, it should be carried out slowly

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\(^{10}\) Please note that ‘valproate’ has been changed to ‘sodium valproate’ to be consistent with the terminology that is being used within this update.

\(^{11}\) Appendix H of the full guideline provides tables for the prognosis for remission of seizures in adults.
(at least 2–3 months) and one drug should be withdrawn at a time. [2004]

1.9.18.5 Particular care should be taken when withdrawing benzodiazepines and barbiturates (may take up to 6 months or longer) because of the possibility of drug-related withdrawal symptoms and/or seizure recurrence. [2004]

1.9.18.6 There should be a failsafe plan agreed with children, young people and adults and their families and/or carers as appropriate, whereby if seizures recur, the last dose reduction is reversed and medical advice is sought. [2004]

1.10 Referral for complex or refractory epilepsy

1.10.1 All children, young people and adults with epilepsy should have access via their specialist to a tertiary service when circumstances require. [2004]

1.10.2 If seizures are not controlled and/or there is diagnostic uncertainty or treatment failure, children, young people and adults should be referred to tertiary services soon for further assessment. Referral should be considered when one or more of the following criteria are present:

- the epilepsy is not controlled with medication within 2 years
- management is unsuccessful after two drugs
- the child is aged under 2 years
- a child, young person or adult experiences, or is at risk of, unacceptable side effects from medication
- there is a unilateral structural lesion
- there is psychological and/or psychiatric co-morbidity
- there is diagnostic doubt as to the nature of the seizures and/or seizure syndrome. [2004]

12 The Guideline Development Group considered that 'soon' meant being seen within 4 weeks.
1.10.3 In children, the diagnosis and management of epilepsy within the first few years of life may be extremely challenging. For this reason, children with suspected epilepsy should be referred to tertiary services early, because of the profound developmental, behavioural and psychological effects that may be associated with continuing seizures. [2004]

1.10.4 Behavioural or developmental regression or inability to identify the epilepsy syndrome in a child, young person or adult should result in immediate referral to tertiary services. [2004]

1.10.5 Children, young people and adults with specific syndromes such as Sturge–Weber syndrome, the hemispheric syndromes, Rasmussen’s encephalitis and hypothalamic hamartoma should be referred to a tertiary epilepsy service. [2004]

1.10.6 Psychiatric co-morbidity and/or negative baseline investigations should not be a contraindication for referral to a tertiary centre. [2004]

1.10.7 The tertiary service should include a multidisciplinary team, experienced in the assessment of children, young people and adults with complex epilepsy, and have adequate access to investigations and treatment by both medical and surgical means. [2004]

1.10.8 The expertise of multidisciplinary teams involved in managing complex epilepsy should include psychology, psychiatry, social work, occupational therapy, counselling, neuroradiology, clinical nurse specialists, neurophysiology, neurology, neurosurgery and neuroanaesthesia. Teams should have MRI and video telemetry facilities available to them. [2004]

1.10.9 The neurosurgeon in the multidisciplinary team should have specialist experience of and/or training in epilepsy surgery and have access to invasive EEG recording facilities. [2004]
1.10.10 Information should be provided to children, young people and adults and families and/or carers as appropriate about the reasons for considering surgery. The benefits and risks of the surgical procedure under consideration should be fully explained before the informed consent is obtained. [2004]

1.11 **Psychological interventions**

1.11.1 Psychological interventions (relaxation, cognitive behaviour therapy, biofeedback) may be used in conjunction with AED therapy in adults where either the person or the specialist considers seizure control to be inadequate with optimal AED therapy. This approach may be associated with an improved quality of life in some people. [2004]

1.11.2 Psychological interventions (relaxation, cognitive behaviour therapy) may be used in children and young people with drug-resistant focal epilepsy. [2004]

1.11.3 Psychological interventions may be used as adjunctive therapy. They have not been proven to affect seizure frequency and are not an alternative to pharmacological treatment. [2004]

1.12 **Ketogenic diet**

1.12.1 Refer children and young people with epilepsy whose seizures have not responded to appropriate AEDs to a tertiary paediatric epilepsy specialist for consideration of the use of a ketogenic diet. [new 2011]

1.12.2 There is currently no good evidence to support the use of the ketogenic diet in adults with epilepsy. [new 2011]
1.13 **Vagus nerve stimulation (VNS)**

1.13.1 Vagus nerve stimulation is indicated for use as an adjunctive therapy in reducing the frequency of seizures in adults who are refractory to antiepileptic medication but who are not suitable for resective surgery. This includes adults whose epileptic disorder is dominated by focal seizures\(^{13}\) (with or without secondary generalisation) or generalised seizures. [2004, amended 2011]

1.13.2 Vagus nerve stimulation is indicated for use as an adjunctive therapy in reducing the frequency of seizures in children and young people who are refractory to antiepileptic medication but who are not suitable for resective surgery. This includes children and young people whose epileptic disorder is dominated by focal seizures\(^{14}\) (with or without secondary generalisation) or generalised seizures\(^{15}\). [2004, amended 2011]

1.14 **Prolonged or repeated seizures and convulsive status epilepticus**

1.14.1 First-line treatment for children, young people and adults with prolonged or repeated generalised, convulsive (tonic–clonic, tonic or clonic) seizures in the community

1.14.1.1 Give immediate emergency care and treatment to children, young people and adults who have prolonged (lasting 5 minutes or more) or repeated (three or more in an hour) convulsive seizures in the community. [2011]

\(^{13}\) In this recommendation, ‘partial seizures’ has been replaced with ‘focal seizures’ to reflect change in terminology since this guideline was published in 2004.

\(^{14}\) In this recommendation, ‘partial seizures’ has been replaced with ‘focal seizures’ to reflect change in terminology since this guideline was published in 2004.

\(^{15}\) Evidence from NICE interventional procedure guidance 50 (March 2004).
1.14.1.2 Administer buccal midazolam* as first-line treatment in children, young people and adults with prolonged or repeated seizures. Administer rectal diazepam* if preferred or if buccal midazolam* is not available. If intravenous access is already established and resuscitation facilities are available, administer intravenous lorazepam. [new 2011]

1.14.1.3 Only prescribe rectal diazepam* or buccal midazolam* for use in the community for children, young people and adults who have had a previous episode of prolonged or serial convulsive seizures. [new 2011]

1.14.1.4 Inform children, young people and adults and their families and/or carers that buccal midazolam* is currently unlicensed. [2011]

1.14.1.5 Treatment should be administered by trained clinical personnel or, if specified by an individually agreed protocol drawn up with the specialist, by family members or carers with appropriate training. [2004]

1.14.1.6 Care must be taken to secure the child’s, young person’s or adult’s airway and assess his or her respiratory and cardiac function. [2004]

1.14.1.7 Depending on response to treatment, the person’s situation and any personalised care plan, call an ambulance, particularly if:

- the seizure is continuing 5 minutes after the emergency medication has been administered
- the person has a history of frequent episodes of serial seizures or has convulsive status epilepticus, or this is the first episode requiring emergency treatment

* At the time of publication (month year), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented in line with normal standards in emergency care.
• there are concerns or difficulties monitoring the person’s airway, breathing, circulation or other vital signs. [new 2011]

1.14.2 Treatment for children, young people and adults with convulsive status epilepticus in hospital

Convulsive status epilepticus

1.14.2.1 For children, young people and adults with ongoing generalised tonic–clonic seizures (convulsive status epilepticus) who are in hospital, immediately:

• secure airway
• give high-concentration oxygen
• assess cardiac and respiratory function
• check blood glucose levels using a finger prick test
• secure intravenous access in a large vein.

See also the suggested protocols in appendix F. [new 2011]

1.14.2.2 Use intravenous lorazepam as first-line treatment in hospital in children, young people and adults with ongoing generalised tonic–clonic seizures (convulsive status epilepticus). Use buccal midazolam* if unable to secure immediate intravenous access. Use a maximum of two doses of the first-line treatment (including pre-hospital treatment). See also the suggested protocols in appendix F. [new 2011]

1.14.2.3 If seizures continue, use 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. [new 2011]

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented in line with normal standards in emergency care.
Refractory convulsive status epilepticus

1.14.2.4 Follow the suggested protocols in appendix F for treating refractory convulsive status epilepticus in secondary care. [2011]

1.14.2.5 Use intravenous midazolam*, propofol* or thiopental sodium* in adults with refractory convulsive status epilepticus. Adequate monitoring, including blood levels of thiopental sodium, and critical life systems support are required. See also the suggested protocols in appendix F. [new 2011]

1.14.2.6 Use intravenous midazolam* or thiopental sodium* in children and young people with refractory convulsive status epilepticus. Adequate monitoring, including blood levels of thiopental sodium, and critical life systems support are required. See also the suggested protocols in appendix F. [2004]

1.14.2.7 As the treatment pathway progresses, the expertise of an anaesthetist/intensivist should be sought. [2004]

1.14.2.8 If either the whole protocol or intensive care is required the tertiary centre should be consulted. [2004]

1.14.2.9 Regular AEDs should be continued at optimal doses and the reasons for status epilepticus should be investigated. [2004]

1.14.2.10 An individual treatment pathway should be formulated for children, young people and adults who have recurrent convulsive status epilepticus. [2004]

1.14.3 Non-convulsive status epilepticus

1.14.3.1 Non-convulsive status epilepticus is uncommon and management is less urgent. A suggested guideline can be found in appendix F. [2004]

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented in line with normal standards in emergency care.
1.15 **Women and girls with epilepsy**

1.15.1 **Information and advice for women and girls with epilepsy**

1.15.1.1 In order to enable informed decisions and choice, and to reduce misunderstandings, women and girls with epilepsy and their partners, as appropriate, must be given accurate information and counselling about contraception, conception, pregnancy, caring for children and breastfeeding, and menopause. [2004]

1.15.1.2 Information about contraception, conception, pregnancy, or menopause should be given to women and girls in advance of sexual activity, pregnancy or menopause, and the information should be tailored to their individual needs. This information should also be given, as needed, to people who are closely involved with women and girls with epilepsy. These may include her family and/or carers. [2004]

1.15.1.3 All healthcare professionals who treat, care for, or support women and girls with epilepsy should be familiar with relevant information and the availability of counselling. [2004]

1.15.1.4 Discuss with women and girls of childbearing potential (including young girls who are likely to need treatment into their childbearing years), and their parents and/or carers if appropriate, the risk of AEDs causing malformations and possible neurodevelopmental delay to an unborn child. Assess the risks and benefits of treatment with individual drugs. There are limited data on risks to the unborn child associated with newer drugs. Specifically discuss the risk of continued use of sodium valproate to the unborn child, being aware that higher doses of sodium valproate (> 800 mg/day) are associated with a greater risk than lower doses (< 800 mg/day). [new 2011]
1.15.1.5 Be aware of the latest data on the risks to the unborn child associated with AED therapy when prescribing for women and girls of present and future childbearing potential. [2011]

1.15.1.6 All women and girls on AEDs should be offered 5 mg per day of folic acid before any possibility of pregnancy. [2004]

1.15.2 Contraception

1.15.2.1 In women of childbearing potential, the possibility of interaction with oral contraceptives should be discussed and an assessment made as to the risks and benefits of treatment with individual drugs. [2004]

1.15.2.2 In girls of childbearing potential, including young girls who are likely to need treatment into their childbearing years, the possibility of interaction with oral contraceptives should be discussed with the child and/or her carer, and an assessment made as to the risks and benefits of treatment with individual drugs. [2004]

1.15.2.3 In women and girls of childbearing potential, the risks and benefits of different contraceptive methods, including hormone-releasing IUDs, should be discussed. [2004]

1.15.2.4 If a woman or girl taking enzyme-inducing AEDs chooses to take the combined oral contraceptive pill, a minimum initial dose of 50 micrograms of oestradiol is recommended. If breakthrough bleeding occurs, the dose of oestradiol should be increased to 75 micrograms or 100 micrograms per day, and ‘tricycling’ (taking three packs without a break) should be considered. [2004]

1.15.2.5 The progestogen-only pill is not recommended as reliable contraception in women and girls taking enzyme-inducing AEDs. [2004, amended 2011]

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16 In this recommendation, ‘progesterone’ has been replaced with ‘progestogen’ to reflect change in terminology since this guideline was published in 2004.
1.15.2.6 The progestogen\textsuperscript{17} implant is not recommended in women and girls taking enzyme-inducing AEDs. [2004, amended 2011]

1.15.2.7 The use of additional barrier methods should be discussed with women and girls taking enzyme-inducing AEDs and oral contraception or having depot injections of progestogen\textsuperscript{18}. [2004]

1.15.2.8 If emergency contraception is required for women and girls taking enzyme-inducing AEDs, the dose of levonorgestrel should be increased to 1.5 mg and 750 micrograms 12 hours apart. [2004]

1.15.2.9 Discuss with women and girls who are taking lamotrigine that the simultaneous use of any oestrogen-based contraceptive can result in a significant reduction of lamotrigine levels and lead to loss of seizure control. When a woman or girl starts or stops taking oral contraceptives, the dose of lamotrigine may need to be adjusted. [new 2011]

1.15.2.10 Refer to the SPC and BNF (available at http://bnf.org) for individual drug advice on the interactions between AEDs and hormonal contraception. [new 2011]

1.15.3 Pregnancy

1.15.3.1 Women and girls with epilepsy need accurate information during pregnancy, and the possibility of status epilepticus and SUDEP should be discussed with all women and girls who plan to stop AED therapy (see section 1.9.18). [2004]

1.15.3.2 All pregnant women and girls with epilepsy should be encouraged to notify their pregnancy, or allow their clinician to notify the pregnancy, to the UK Epilepsy and Pregnancy Register (www.epilepsyandpregnancy.co.uk). [2004]

\textsuperscript{17} In this recommendation, ‘progesterone’ has been replaced with ‘progestogen’ to reflect change in terminology since this guideline was published in 2004.

\textsuperscript{18} In this recommendation, ‘progesterone’ has been replaced with ‘progestogen’ to reflect change in terminology since this guideline was published in 2004.
1.15.3.3 The clinician should discuss with the woman and girl the relative benefits and risks of adjusting medication to enable her to make an informed decision. Where appropriate, the woman’s or girl’s specialist should be consulted. [2004]

1.15.3.4 Women and girls with generalised tonic–clonic seizures should be informed that the fetus may be at relatively higher risk of harm during a seizure, although the absolute risk remains very low, and the level of risk may depend on seizure frequency. [2004]

1.15.3.5 Women and girls should be reassured that there is no evidence that focal¹⁹, absence and myoclonic seizures affect the pregnancy or developing fetus adversely unless they fall and sustain an injury. [2004, amended 2011]

1.15.3.6 Women and girls should be reassured that an increase in seizure frequency is generally unlikely in pregnancy or in the first few months after birth. [2004]

1.15.3.7 Generally, women and girls may be reassured that the risk of a tonic–clonic seizure during the labour and the 24 hours after birth is low (1–4%). [2004]

1.15.3.8 Women and girls with epilepsy should be informed that although they are likely to have healthy pregnancies, their risk of complications during pregnancy and labour is higher than for women and girls without epilepsy. [2004]

1.15.3.9 Care of pregnant women and girls should be shared between the obstetrician and the specialist. [2004]

1.15.3.10 Pregnant women and girls who are taking AEDs should be offered a high-resolution ultrasound scan to screen for structural anomalies. This scan should be performed at 18–20 weeks’ gestation by an appropriately trained ultrasonographer, but earlier

¹⁹ In this recommendation, ‘partial seizures’ has been replaced with ‘focal seizures’ to reflect change in terminology since this guideline was published in 2004.
scanning may allow major malformations to be detected sooner. [2004]

1.15.3.11 The risk of seizures during labour is low, but it is sufficient to warrant the recommendation that delivery should take place in an obstetric unit with facilities for maternal and neonatal resuscitation and treating maternal seizures. [2004]

1.15.3.12 All children born to mothers taking enzyme-inducing AEDs should be given 1 mg of vitamin K parenterally at delivery. [2004]

1.15.3.13 Genetic counselling should be considered if one partner has epilepsy, particularly if the partner has idiopathic epilepsy and a positive family history of epilepsy. [2004]

1.15.3.14 Although there is an increased risk of seizures in children of parents with epilepsy, children, young people and adults with epilepsy should be given information that the probability that a child will be affected is generally low. However, this will depend on the family history. [2004]

1.15.3.15 Advanced planning, including the development of local protocols for care, should be implemented in obstetric units that deliver babies of women and girls with epilepsy. [2004]

1.15.3.16 Joint epilepsy and obstetric clinics may be convenient for mothers and healthcare professionals but there is insufficient evidence to recommend their routine use. [2004]

1.15.3.17 It is, however, important that there should be regular follow-up, planning of delivery, and liaison between the specialist or epilepsy team and the obstetrician or midwife. [2004]

1.15.3.18 Aim for seizure freedom before conception and during pregnancy (particularly for women and girls with generalised tonic–clonic seizures) but consider the risk of adverse effects of AEDs and use the lowest effective dose of each AED. [new 2011]
1.15.3.19 Do not routinely monitor AED levels during pregnancy. If seizures increase or are likely to increase, monitoring AED levels (particularly levels of lamotrigine and phenytoin which may be particularly affected in pregnancy) may be useful when making dose adjustments. [new 2011]

1.15.4 Breastfeeding

1.15.4.1 All women and girls with epilepsy should be encouraged to breastfeed, except in very rare circumstances. Breastfeeding for most women and girls taking AEDs is generally safe and should be encouraged. However, each mother needs to be supported in the choice of feeding method that best suits her and her family. [2004]

1.15.4.2 Prescribers should consult individual drug advice in the SPC and the BNF (available at http://bnf.org)\(^\text{20}\) when prescribing AEDs for women and girls who are breastfeeding. The decision regarding AED therapy and breastfeeding should be made between the woman or girl and the prescriber, and be based on the risks and benefits of breastfeeding against the potential risks of the drug affecting the child. [2004, amended 2011]

1.15.5 After the birth

1.15.5.1 Parents of new babies or young children should be informed that introducing a few simple safety precautions may significantly reduce the risk of accidents and minimise anxiety. An approaching birth can be an ideal opportunity to review and consider the best and most helpful measures to start to ensure maximum safety for both mother and baby. [2004]

\(^{20}\) In this recommendation, the original referral to appendix 5 of the BNF has been removed and replaced with more up-to-date source reference material because this appendix no longer exists and has therefore become obsolete since this guideline was published in 2004.
1.15.5.2 Information should be given to all parents about safety precautions to be taken when caring for the baby (see appendix D\textsuperscript{21} of the full guideline). [2004]

1.15.5.3 Parents should be reassured that the risk of injury to the infant caused by maternal seizure is low. [2004]

1.16 **Children, young people and adults with learning disabilities (see also sections 1.15 and 1.17)**

1.16.1 **Diagnosis (see also section 1.5)**

1.16.1.1 It can be difficult to diagnose epilepsy in children, young people and adults with learning disabilities, and so care should be taken to obtain a full clinical history. Confusion may arise between stereotypic or other behaviours and seizure activity. [2004]

1.16.1.2 It is important to have an eye witness account supplemented by corroborative evidence (for example, a video account), where possible. [2004]

1.16.1.3 Clear, unbiased reporting is essential. Witnesses may need education to describe their observations accurately. [2004]

1.16.2 **Investigations (see also section 1.6)**

1.16.2.1 Those with learning disabilities may require particular care and attention to tolerate investigations. [2004]

1.16.2.2 Facilities should be available for imaging under anaesthesia, if necessary. [2004]

1.16.2.3 In the child or young person presenting with epilepsy and learning disability, investigations directed at determining an underlying cause should be undertaken. [2004]

\textsuperscript{21} Appendix D of the full guideline provides a checklist for the information needs of women and girls with epilepsy, and practical information for mothers with epilepsy.
1.16.3 Management (see also section 1.8)

1.16.3.1 Enable children, young people and adults who have learning disabilities, and their family and/or carers where appropriate, to take an active part in developing a personalised care plan for treating their epilepsy while taking into account any comorbidities. [new 2011]

1.16.3.2 Ensure adequate time for consultation to achieve effective management of epilepsy in children, young people and adults with learning disabilities. [new 2011]

1.16.3.3 In making a care plan for a child, young person or adult with learning disabilities and epilepsy, particular attention should be paid to the possibility of adverse cognitive and behavioural effects of AED therapy. [2004]

1.16.3.4 The recommendations on choice of treatment and the importance of regular monitoring of effectiveness and tolerability are the same for those with learning disabilities as for the general population. [2004]

1.16.3.5 Do not discriminate against children, young people and adults with learning disabilities and offer the same services, investigations and therapies as for the general population. [new 2011]

1.16.3.6 Every therapeutic option should be explored in children, young people and adults with epilepsy in the presence or absence of learning disabilities. [2004]

1.16.3.7 Healthcare professionals should be aware of the higher risks of mortality for children, young people and adults with learning disabilities and epilepsy and discuss these with them, their families and/or carers. [2004]

1.16.3.8 All children, young people and adults with epilepsy and learning disabilities should have a risk assessment including:
1.17 Young people with epilepsy (see also section 1.15)

1.17.1 The physical, psychological and social needs of young people with epilepsy should always be considered by healthcare professionals. Attention should be paid to their relationships with family and friends, and at school. [2004]

1.17.2 Healthcare professionals should adopt a consulting style that allows the young person with epilepsy to participate as a partner in the consultation. [2004]

1.17.3 Decisions about medication and lifestyle issues should draw on both the expertise of the healthcare professional and the experiences, beliefs and wishes of the young person with epilepsy as well as their family and/or carers. [2004]

1.17.4 During adolescence a named clinician should assume responsibility for the ongoing management of the young person with epilepsy and ensure smooth transition of care to adult services, and be aware of the need for continuing multi-agency support. [2004]

1.17.5 Multidisciplinary services provided jointly by adult and paediatric specialists have a key role in the care of the young person with epilepsy. This can facilitate the transition from paediatric to adult services and aid in the dissemination of information. [2004]
1.17.6 Before the transition to adult services is made, diagnosis and management should be reviewed and access to voluntary organisations, such as support groups and epilepsy charities, should be facilitated. [2004]

1.17.7 The information given to young people should cover epilepsy in general and its diagnosis and treatment, the impact of seizures and adequate seizure control, treatment options including side effects and risks, and the risks of injury. Other important issues to be covered are the possible consequences of epilepsy on lifestyle and future career opportunities and decisions, driving and insurance issues, social security and welfare benefit issues, sudden death and the importance of adherence to medication regimes. Information on lifestyle issues should cover recreational drugs, alcohol, sexual activity and sleep deprivation (see section 1.3). [2004]

1.17.8 The diagnosis and management of epilepsy should be reviewed during adolescence. [2004]

1.18 Older people with epilepsy

1.18.1 Do not discriminate against older people and offer the same services, investigations and therapies as for the general population. [new 2011]

1.18.2 Pay particular attention to pharmacokinetic and pharmacodynamic issues with polypharmacy and comorbidity in older people with epilepsy. Consider using lower doses of AEDs and, if using carbamazepine, offer controlled-release carbamazepine preparations. [new 2011]
1.19 **Children, young people and adults from black and minority ethnic groups**

1.19.1 Children, young people and adults from black and minority ethnic groups may have different cultural and communication needs and these should be considered during diagnosis and management. The need for interpretation should be considered alongside other means of ensuring that a person’s needs are appropriately met. [2004]

1.19.2 An interpreter should have both cultural and medical knowledge. Interpreters from the family are generally not suitable because of issues such as confidentiality, privacy, personal dignity, and accuracy of translation. [2004]

1.19.3 Information, including information about employment rights and driving, should be available in an appropriate format or through other appropriate means for children, young people and adults who do not speak or read English. [2004]

1.20 **Review**

1.20.1 Children, young people and adults with epilepsy should have a regular structured review and be registered with a general medical practice. [2004]

1.20.2 Adults should have a regular structured review with their GP, but depending on the person’s wishes, circumstances and epilepsy, the review may be carried out by the specialist. [2004]

1.20.3 Children and young people should have a regular structured review with a specialist. [2004]

1.20.4 For adults, the maximum interval between reviews should be 1 year but the frequency of review will be determined by the person’s epilepsy and their wishes. [2004]
1.20.5 For children and young people, the maximum interval between reviews should be 1 year, but the frequency of reviews should be determined by the child’s or young person’s epilepsy and their wishes and those of the family and/or carers. The interval between reviews should be agreed between the child or young person, their family and/or carers as appropriate, and the specialist, but is likely to be between 3 and 12 months. [2004]

1.20.6 Adults should have regular reviews. In addition, access to either secondary or tertiary care should be available to ensure appropriate diagnosis, investigation and treatment if the person or clinician view the epilepsy as inadequately controlled. [2004]

1.20.7 Adults with well-controlled epilepsy may have specific medical or lifestyle issues (for example, pregnancy or drug cessation) that may need the advice of a specialist. [2004]

1.20.8 If the structured review is to be conducted by the specialist, this may be best provided in the context of a specialist clinic. [2004]

1.20.9 Treatment should be reviewed at regular intervals to ensure that children, young people and adults with epilepsy are not maintained for long periods on treatment that is ineffective or poorly tolerated and that concordance with prescribed medication is maintained. [2004]

1.20.10 Annual review should include an enquiry about side effects and a discussion of the treatment plan to ensure concordance and adherence to medication. [2004]

1.20.11 At the review, children, young people and adults should have access to: written and visual information; counselling services; information about voluntary organisations; epilepsy specialist nurses; timely and appropriate investigations; referral to tertiary services including surgery, where appropriate. [2004]
2 Notes on the scope of the guidance

NICE guidelines are developed in accordance with a scope that defines what
the guideline will and will not cover. The scope of this guideline is available
from www.nice.org.uk/[NICE to add details].

The guideline addresses the diagnosis, treatment and management of
epilepsy in children, young people and adults. It does not cover the diagnosis,
treatment or management of epilepsy in neonates or the diagnosis or
management of febrile convulsions.

The guideline makes recommendations concerning the care provided by
healthcare professionals who have direct contact with, or make decisions
concerning, the care of people with epilepsy. It deals with care in primary,
secondary and tertiary centres, and integrated care for epilepsy may span all
these sectors. The delivery of tertiary procedures, such as surgical
techniques, is not included. The guideline will also be relevant to, but does not
cover the practice of, those working in the occupational health services, social
services, educational services or the voluntary sector.

In 2011 the pharmacological management sections of the guideline were
updated.
How this guideline was developed

NICE commissioned the National Clinical Guideline Centre to develop this guideline. The Centre established a Guideline Development Group (see appendix A), which reviewed the evidence and developed the recommendations. An independent Guideline Review Panel oversaw the development of the guideline (see appendix B).

There is more information about how NICE clinical guidelines are developed on the NICE website (www.nice.org.uk/HowWeWork). A booklet, ‘How NICE clinical guidelines are developed: an overview for stakeholders, the public and the NHS’ (fourth edition, published 2009), is available from NICE publications (phone 0845 003 7783 or email publications@nice.org.uk and quote reference N1739).

3 Implementation

NICE has developed tools to help organisations implement this guidance (see www.nice.org.uk/guidance/CG[XX]).

4 Research recommendations

The Guideline Development Group has made the following recommendations for research, based on its review of evidence, to improve NICE guidance and patient care in the future.

4.1 Newly diagnosed seizures (focal and generalised) – monotherapy

How do the newer AEDs compare in efficacy to the standard AEDs in the treatment of newly diagnosed epilepsy?

- Focal seizures: carbamazepine, eslicarbazepine acetate, lamotrigine, lacosamide, levetiracetam, pregabalin and zonisamide.
- Generalised seizures: lamotrigine, levetiracetam, sodium valproate and zonisamide.
**Why this is important**

Levetiracetam and other AEDs licensed for the treatment of focal and generalised seizures since publication of the original guideline ‘The epilepsies’ (NICE clinical guideline 20) in 2004 have not been evaluated as first-line monotherapy.

The research should include:

- a prospective randomised controlled trial
- all age groups
- subgroup analyses on seizure types and syndromes
- primary outcome of seizure freedom
- secondary outcomes, including seizure reduction, quality of life and cognitive outcome
- an attempt to obtain data on pharmaco-resistance.

**4.2 Epilepsy syndromes**

What is the initial and add-on AEDs of choice in the treatment of the epilepsy syndromes with onset in childhood, for example, myoclonic-astatic epilepsy and Dravet syndrome (SMEI)?

**Why this is important**

Despite the need to diagnose individual epilepsy syndromes, there is little evidence on the most appropriate initial or add-on AEDs in the treatment of the rarer epilepsies.

The research should include:

- multicentre randomised controlled comparative trials with centralised national data collection
- the ketogenic diet as one of the randomised treatments
- primary outcome of seizure freedom
- secondary outcomes, including seizure reduction, quality of life and cognitive outcome
- an attempt to obtain data on pharmaco-resistance
• the possibility of including all children with specific epilepsy syndromes for consideration in the trial.

4.3 Infantile spasms

Does treatment response relate to cause in infantile spasms? Does early treatment success in seizure control and resolution of the hypsarrhythmic EEG influence the long-term developmental and cognitive outcomes more than the underlying cause of the spasms?

Why this is important

The UK Infantile Spasms Study (UKISS)\textsuperscript{22} demonstrated 14-day outcome efficacy of steroids over vigabatrin, although this excluded children with tuberous sclerosis. This study provided no specific sub-group analysis based on the cause of the spasms. There was no analysis on the effect of treatment lag (delay) on the study findings. Further data are available on behavioural outcomes at 14 months and 4 years with regard to different treatments but with no analysis based on cause or treatment lag. Further developmental and cognitive outcomes would be useful, including response by specific cause and by treatment lag.

The research should include:

• prospective randomised design, including sub-group analyses based on both cause and treatment lag; this would require large numbers of patients and would need to be multicentre, possibly involving Western Europe

• EEG outcomes

• developmental status at presentation, and at follow-up

• an attempt to obtain data on pharmaco-resistance.

4.4 Treatment of convulsive status epilepticus (that is, not just refractory)

What is the most effective and safest anticonvulsant to treat:

- established (usually > 30 min) convulsive status epilepticus
- refractory convulsive status epilepticus?

Why is this important?
Convulsive status epilepticus should be treated as an emergency. The most important aspect of treatment is to try to stop the seizure. The medication currently used first is phenytoin, however this has to be used with care with close monitoring because of the risk of cardiac arrhythmia. There are alternatives (sodium valproate and levetiracetam) for which there are few comparative data. Prompt and successful treatment of convulsive status epilepticus will in many cases prevent the need for admission to an intensive care unit. Convulsive status epilepticus that is refractory to first-line treatment is rare and often complicated by irreversible neurological and intellectual sequelae, including death. Reasons for these complications include the underlying cause of refractory convulsive status epilepticus and its duration, but also its management. Most patients with refractory convulsive status epilepticus will be managed on an intensive care unit. There are no agreed drugs or treatment protocols for treating refractory convulsive status epilepticus. The three most commonly used anticonvulsants are thiopental sodium, midazolam and propofol (although propofol is rarely used in children).

There are very limited data on the treatment of refractory convulsive status epilepticus in adults, children and young people. A 2-year audit of all children and young people with refractory convulsive status epilepticus treated on an intensive care unit in England, Wales and Scotland has recently been completed. This will provide unique epidemiological data on paediatric refractory convulsive status epilepticus and its current management. These data will facilitate the design of a randomised controlled trial of specific drug treatments and protocols.
The research should include:

- a multicentre randomised comparative trial of intravenous levetiracetam, sodium valproate and phenytoin as initial treatment of convulsive status epilepticus
- a multicentre, randomised controlled trial of treatment of refractory convulsive status epilepticus on intensive care units to include midazolam and thiopental sodium (and propofol in adults)
- primary outcome should be cessation of convulsive status epilepticus
- secondary outcomes should include a recurrence within a designated period (12 hours), mortality and morbidity
- cost data should include treatment costs and days on intensive care.

### 4.5 AEDs and pregnancy

What is the malformation rate and longer term neurodevelopmental outcome of children born to mothers who have taken AEDs in pregnancy?

**Why this is important**

Pregnancy registers are increasing the data that are available on established AEDs; however, these registers may give malformation rates but do not provide controlled long-term data on neurodevelopmental outcome.

The research should include:

- measures of maternal outcome, including seizure frequency and quality of life
- major and minor rates of congenital malformations
- prospective neurodevelopmental (including cognitive) and behavioural outcomes in children born to women and girls with epilepsy (these should be undertaken on a long-term basis and ideally using a cohort study, followed from birth and until adult life).
5 Other versions of this guideline

5.1 Full guideline

The full guideline, ‘The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care (a partial update of clinical guideline 20)’, contains details of the methods and evidence used to develop the guideline. It is published by the National Clinical Guideline Centre, and is available from our website (www.nice.org.uk/guidance/CG[XX]/FullGuidance).

5.2 Quick reference guide

A quick reference guide for healthcare professionals is available from www.nice.org.uk/guidance/CG[XX]/QuickRefGuide

For printed copies, phone NICE publications on 0845 003 7783 or email publications@nice.org.uk (quote reference number N[XXXX]).

5.3 ‘Understanding NICE guidance’

A summary for patients and carers (‘Understanding NICE guidance’) is available from www.nice.org.uk/guidance/CG[XX]/PublicInfo

For printed copies, phone NICE publications on 0845 003 7783 or email publications@nice.org.uk (quote reference number N[XXXX]).

We encourage NHS and voluntary sector organisations to use text from this booklet in their own information about epilepsy.

6 Related NICE guidance

Published


7 Updating the guideline

NICE clinical guidelines are updated so that recommendations take into account important new information. New evidence is checked 3 years after publication, and healthcare professionals and patients are asked for their views; we use this information to decide whether all or part of a guideline needs updating. If important new evidence is published at other times, we may decide to do a more rapid update of some recommendations. Please see our website for information about updating the guideline.
Appendix A: The Guideline Development Group, National Clinical Guideline Centre and NICE project team

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For this guideline, the Guideline Development Group was assisted by a number of co-opted experts, who were chosen because of their knowledge in a particular area.

2011 Guideline (partial update)

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Michelle Wallwin
Editor
Appendix B: The Guideline Review Panel

The Guideline Review Panel is an independent panel that oversees the development of the guideline and takes responsibility for monitoring adherence to NICE guideline development processes. In particular, the panel ensures that stakeholder comments have been adequately considered and responded to. The panel includes members from the following perspectives: primary care, secondary care, lay, public health and industry.

2011 Guideline (partial update)
To be completed

2004 Guideline

Mr Barry Stables
Patient member

Dr Imogen Stephens
Joint Director of Public Health, Western Sussex Primary Care Trust

Dr Kevork Hopayian
GP, Suffolk

Professor Mike Drummond (Chair)
Director, Centre for Health Economics, University of York

Dr Robert Walker
Clinical Director, West Cumbria Primary Care
Appendix C: Outline care algorithms

**Adults**

1. Suspected seizure
2. Primary care
3. Information obtained about the event
   - Physical examination
4. A&E
   - (protocols in place for assessment)
   - Initial screening by physician
5. Diagnostic doubt
6. Suspected epileptic seizure
7. Referral to specialist as soon as possible
   - (The GDG recommended within 2 weeks)
8. Diagnosis by specialist with investigations as necessary
9. Uncertain
10. Epilepsy
11. Non-epileptic attack disorder
12. Referral to psychological or psychiatric services
13. Investigation and classification by seizure type and epilepsy syndrome by specialist
14. Treatment
15. Referral to tertiary care
16. Special groups
   - People with learning disabilities
   - Black and ethnic minority groups
   - Older people
17. Women with epilepsy
18. Prolonged or repeated seizures
   - Status epilepticus
19. Regular structured review for all

**KEY:** ——— As necessary
Children and young people

Suspected seizure

Primary care

Information obtained about the event
Physical examination

A&E
(protocols in place for assessment)
Initial screening by paediatrician

Diagnostic doubt

Referral to specialist as soon as possible
(The GDG recommended within 2 weeks)

Diagnosis by specialist with investigations as necessary
(see box A)

Uncertain
Further investigation, including assessment of other physical causes (e.g. cardiac) or Referral to tertiary care (see box A)

Epilepsy

Investigation and classification by seizure type and epilepsy syndrome by specialist (see box A)

Treatment
(see box A)

Non-epileptic attack disorder

Referral to psychological or psychiatric services

Special groups
- Children and young people with learning disabilities
- Black and ethnic minority groups
- Young people with epilepsy (see box A)

Young women and girls with epilepsy
(see box A)

Prolonged or repeated seizures
Status epilepticus
(see box A)

Regular structured review for all (see box A)

KEY: ———— As necessary
## Box A Cross reference for algorithms

<table>
<thead>
<tr>
<th>Section</th>
<th>Page number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Treatment with AEDs only in exceptional circumstances</td>
<td>24</td>
</tr>
<tr>
<td>Diagnosis and investigations</td>
<td>16–17</td>
</tr>
<tr>
<td>Further investigation</td>
<td>17–21</td>
</tr>
<tr>
<td>Investigation and classification by seizure type and epilepsy syndrome</td>
<td>17–21</td>
</tr>
<tr>
<td>Referral to tertiary care</td>
<td>38–40</td>
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<tr>
<td>Treatment</td>
<td>22–38</td>
</tr>
<tr>
<td>Prolonged or repeated seizures; status epilepticus</td>
<td>41–44</td>
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<tr>
<td>Women or girls with epilepsy</td>
<td>45–51</td>
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<tr>
<td>Special groups</td>
<td>51–55</td>
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<tr>
<td>Regular structured review</td>
<td>55–56</td>
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<tr>
<td>Appropriate information</td>
<td>12–14</td>
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</table>
Appendix D: Differential diagnosis of epilepsy in children, young people and adults

Differential diagnosis of epilepsy in adults
Abnormal movements predominate

- Generalised convulsive movements
- Drop attacks
- Transient focal motor attacks
- Facial muscle and eye movements
- Episodic phenomena in sleep

Disturbed awareness, thoughts, and sensations predominate

- Loss of awareness
- Transient focal sensory attacks
- Psychic experiences
- Aggressive or vocal outbursts
- Prolonged confusional or fugue states

- Epilepsy
- Syncope with secondary jerking movements
- Primary cardiac or respiratory abnormalities, presenting with secondary anoxic seizures
- Involuntary movement disorders and other neurological conditions
- Hyperekplexia
- Non-epileptic attack disorder (NEAD)

- Epilepsy
- Cardiovascular
- Movement disorders
- Brainstem, spinal, or lower limb abnormalities
- Catechol
- Metabolic disorders
- Isotonic drop attacks
- Vertebrobasilar ischaemia

- Focal motor seizures
- Tic
- Transient cerebral ischaemia
- Tonic spasms of multiple sclerosis
- Paroxysmal movement disorders

- Partial seizures
- Movement disorders
- Other neurological disorders

- Normal physiological movements
- Frontal lobe epilepsy
- Other epilepsy
- Pathological fragmentary myoclonus
- Restless leg syndrome
- Non-REM/REM parasomnias
- Sleep apnoea
- Other movements in sleep

- Somatosensory attacks: epileptic seizure, transient ischaemic attack, hyperventilation
- Transient vestibular symptoms: peripheral vestibular disease, epilepsy
- Visual symptoms: migraine, transient ischaemic attack, epilepsy

- Epilepsy
- Migraine
- Panic attacks
- Drug induced flashbacks
- Hallucinations or illusions caused by loss of a primary sense
- Psychotic hallucinations and delusions
- Non-epileptic attack disorder (NEAD)

- Related to learning disability
- Epilepsy
- Volitional

- Acute encephalopathy
- Non-convulsive status epilepticus
- Intermittent psychosis
- Transient global amnesia
- Hysterical fugue
Differential diagnosis of epilepsy in children and young people
**History of Event / Attack**
- Frequency
- Timing
- Triggers
- Warning beforehand
- Colour change
- Alteration in conscious level
- Motor phenomena
- Duration of attack
- Symptoms following attack

**What is the trigger for the attack?**
- Only during sleep?
- Related to feeding?
- With a fever?
- On initiation of movement?
- With excitement/emotion?
- Following unpleasant/painful stimuli?
- Boredom/concentration

**What is the colour change?**
- Pallor
- Cyanosis
- Flushing
- Structural cardiac lesion
- Cyanotic breath-holding attack
- Gastro-oesophageal reflux

**What is the predominant motor phenomenon?**
- Repetitive stereotyped spasm?
- Hypertonia?
- Hypotonia (include FALLS)?
- Dystonia?
- Unsteadiness?

**What is the trigger for the attack?**
- Cardiac arrhythmias
- Structural cardiac lesion
- Benign myoclonus of infancy
- Paroxysmal dystonia
- Sandifer syndrome/GOR
- Benign paroxysmal torticollis
- Alternating hemiplegia
- Infantile spasms
- Self gratification behaviour
- Shuddering attacks
- Benign sleep myoclonus

**TODDLER**
- Cardiac arrhythmias
- Reflex anoxic seizures
- Cyanotic breath-holding attacks
- Hyperexplexia
- Myoclonus
- Paroxysmal dyskinesias
- Sandifer syndrome
- Benign paroxysmal vertigo/torticollis
- Migraine
- Cataplexy
- Akineic (drop) attacks
- Febrile convulsions
- Overflow movements
- Self gratification behaviour
- Stereotypies/stylised behaviour (eg. Children with learning difficulties)
- Head banging
- Confusional arousal
- Night terrors

**OLDER CHILD**
- Cardiac arrhythmias
- Neurocardiogenic syncope
- Reflex anoxic seizures
- Neurocardiogenic syncope
- Hyperexplexia
- Myoclonus
- TIAs
- Paroxysmal dyskinesias
- Benign paroxysmal vertigo/torticollis
- Migraine
- Eye movement disorders
- Episodic ataxia
- Cataplexy
- Akineic (drop) attacks
- Day dreams
- Hyperventilation/panic attacks
- Non epileptic attack disorder
- Pseudo-syncpe or psychogenic syncope
- Stereotypies/stylised behaviour (eg. Children with learning difficulties)
- Confusional arousal
- REM sleep disorders
- Night terrors
Appendix E: Pharmacological treatment

The tables that follow provide a summary reference guide to pharmacological treatment. They were updated in 2011. Licensing details are listed under table 3. All drugs are listed in alphabetical order.
## Table 1 AED options by seizure type

<table>
<thead>
<tr>
<th>Seizure type</th>
<th>First-line AEDs</th>
<th>Adjunctive AEDs</th>
<th>Other AEDs that may be considered</th>
<th>Do not offer AEDs (may worsen seizures)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Generalised tonic–clonic</td>
<td>Carbamazepine</td>
<td>Clobazam*</td>
<td>Lamotrigine</td>
<td>(if there are absence or myoclonic seizures, or if JME suspected)</td>
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<tr>
<td></td>
<td>Lamotrigine*</td>
<td>Levetiracetam</td>
<td>Sodium valproate</td>
<td>Carbamazepine</td>
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<tr>
<td></td>
<td>Oxcarbazepine*</td>
<td>Sodium valproate</td>
<td>Topiramate*</td>
<td>Gabapentin</td>
</tr>
<tr>
<td></td>
<td>Sodium valproate</td>
<td></td>
<td></td>
<td>Oxcarbazepine</td>
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<td>Topiramate</td>
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<td>Phenytoin</td>
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<td></td>
<td>Vigabatrin</td>
</tr>
<tr>
<td>Tonic or atonic</td>
<td>Sodium valproate</td>
<td>Lamotrigine*</td>
<td>Rufinamide</td>
<td>Carbamazepine</td>
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<td>Gabapentin</td>
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<td>Absence</td>
<td>Ethosuximide</td>
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<td>Clonazepam</td>
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<td>Vigabatrin</td>
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<td>Focal with/without secondary generalisation</td>
<td>Carbamazepine</td>
<td>Carbamazepine</td>
<td>Eslicarbazepine acetate*</td>
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<td>Topiramate</td>
<td>Zonisamide*</td>
<td>Vigabatrin</td>
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* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
## Table 2 AED options by epilepsy syndrome

<table>
<thead>
<tr>
<th>Epilepsy syndrome</th>
<th>First-line AEDs</th>
<th>Adjunctive AEDs</th>
<th>Other AEDs</th>
<th>Do not offer AEDs (may worsen seizures)</th>
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<td>Childhood absence epilepsy</td>
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<td>Vigabatrin</td>
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<td>Juvenile absence epilepsy</td>
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<td>Epilepsy with generalised tonic–clonic seizures only</td>
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<td>Carbamazepine Lamotrigine Levetiracetam Sodium Valproate Topiramate</td>
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<td>Idiopathic Generalised Epilepsies</td>
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<td></td>
<td></td>
<td></td>
<td>Vigabatrin</td>
<td></td>
</tr>
<tr>
<td>Infantile spasms</td>
<td>Steroid or (prednisolone or tetracosactide*) vigabatrin (when infantile spasms not due to tuberous)</td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>First-line treatment</th>
<th>Second-line treatment</th>
<th>Third-line treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign epilepsy with centrotemporal spikes</td>
<td>Carbamazepine, Lamotrigine, Levetiracetam, Oxcarbazepine, Sodium valproate</td>
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<tr>
<td>Panyiotopoulos syndrome and late-onset childhood occipital epilepsy (Gastaut type)</td>
<td>Carbamazepine* Lamotrigine* Levetiracetam* Oxcarbazepine* Sodium valproate</td>
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<tr>
<td>Dravet syndrome</td>
<td>Sodium valproate, Topiramate* Clobazam* Stiripentol</td>
<td></td>
<td>Carbamazepine Gabapentin Lamotrigine Oxcarbazepine Phenytoin Pregabalin Tiagabine Vigabatrin</td>
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<tr>
<td>Continuous spike wave of slow sleep</td>
<td>Referral to a tertiary epilepsy specialist</td>
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<tr>
<td>Lennox–Gastaut syndrome</td>
<td>Sodium valproate, Lamotrigine Rufinamide Topiramate Felbamate *</td>
<td></td>
<td>Carbamazepine Gabapentin Oxcarbazepine Pregabalin Tiagabine Vigabatrin</td>
</tr>
<tr>
<td>Landau–Kleffner syndrome</td>
<td>Referral to a tertiary epilepsy specialist</td>
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<td></td>
</tr>
<tr>
<td>Myoclonic-astatic epilepsy</td>
<td>Referral to a tertiary epilepsy specialist</td>
<td></td>
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</tr>
</tbody>
</table>

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented.
Licensing indications

Detailed below are drugs that have been recommended but which do not currently have licensed indications for these seizures types or syndromes or particular populations.

Table 3 Licensing indications of the guideline AEDs

<table>
<thead>
<tr>
<th>Seizure type/syndrome</th>
<th>Drug</th>
<th>Details of licensing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Treatment of refractory focal seizures</td>
<td>Clobazam</td>
<td>At the time of publication, clobazam did not have UK marketing authorisation for use in children younger than 3 years (BNFC). This was because of insufficient experience of the use of this drug in children younger than 3 years to enable any dosage recommendation to be made (SPC).</td>
</tr>
<tr>
<td></td>
<td>Gabapentin</td>
<td>At the time of publication, gabapentin did not have UK marketing authorisation for use in children younger than 6 years and at doses over 50 mg/kg daily in children younger than 12 years (BNFC). The use of gabapentin was not recommended in this age group owing to the lack of sufficient supporting data (SPC).</td>
</tr>
<tr>
<td></td>
<td>Eslicarbazepine acetate</td>
<td>At the time of publication, eslicarbazepine acetate did not have UK marketing authorisation for use in children younger than 18 years. It was not recommended owing to a lack of data on safety and efficacy (SPC).</td>
</tr>
<tr>
<td></td>
<td>Pregabalin</td>
<td>At the time of publication, pregabalin did not have UK marketing authorisation for use in children (BNF). Pregabalin was not recommended for use in children younger than 12 years and adolescents (12–17 years) owing to insufficient data on safety and efficacy (SPC).</td>
</tr>
<tr>
<td></td>
<td>Zonisamide</td>
<td>At the time of publication, zonisamide did not have UK marketing authorisation for use in children younger than 18 years owing to insufficient data on safety and efficacy (SPC).</td>
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<tr>
<td>Category</td>
<td>Drug</td>
<td>Information</td>
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<td>GTC</td>
<td>Oxcarbazepine</td>
<td>At the time of publication, oxcarbazepine did not have UK marketing authorisation for GTC seizures (BNF). It had authorisation for focal with or without secondarily generalised tonic–clonic seizures (BNF).</td>
</tr>
<tr>
<td></td>
<td>Clobazam</td>
<td>At the time of publication, clobazam did not have UK marketing authorisation for use in children younger than 3 years (BNFC). There was insufficient experience of the use of this drug in children younger than 3 years to enable any dosage recommendation to be made (SPC).</td>
</tr>
<tr>
<td>Absence seizures</td>
<td>Clobazam</td>
<td>At the time of publication, clobazam did not have UK marketing authorisation for use in children younger than 3 years (BNFC). This was because of insufficient experience of the use of this drug in children younger than 3 years to enable any dosage recommendation to be made (SPC).</td>
</tr>
<tr>
<td></td>
<td>Lamotrigine</td>
<td>At the time of publication, lamotrigine had UK marketing authorisation for monotherapy of typical absence seizures for those aged 2–12 years only. There was not authorisation outside of this age range (BNF).</td>
</tr>
<tr>
<td></td>
<td>Levetiracetam</td>
<td>At the time of publication, levetiracetam did not have UK marketing authorisation for use in absence seizures but had authorisation for focal seizures with or without secondary generalisation and adjunctive therapy for myoclonic and GTC seizures (BNFC).</td>
</tr>
<tr>
<td></td>
<td>Topiramate</td>
<td>At the time of publication, topiramate did not have UK marketing authorisation for use in absence seizures but had authorisation for focal seizures, GTC seizures and seizures associated with Lennox–Gastaut syndrome (BNF).</td>
</tr>
<tr>
<td>Seizure Type</td>
<td>Drug</td>
<td>Status</td>
</tr>
<tr>
<td>----------------------</td>
<td>---------------</td>
<td>--------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Absence seizures</td>
<td>Zonisamide</td>
<td>At the time of publication, zonisamide did not have UK marketing authorisation for use in absence seizures but had authorisation for adjunctive therapy for adult patients with partial seizures, with or without secondary generalisation (BNF).</td>
</tr>
<tr>
<td>Myoclonic seizures</td>
<td>Clobazam</td>
<td>At the time of publication, clobazam did not have UK marketing authorisation for use in children younger than 3 years (BNFC). This was because of insufficient experience of the use of this drug in children younger than 3 years to enable any dosage recommendation to be made (SPC).</td>
</tr>
<tr>
<td></td>
<td>Levetiracetam</td>
<td>At the time of publication, levetiracetam did not have UK marketing authorisation for monotherapy use in myoclonic seizures but had authorisation for monotherapy and adjunctive treatment of focal seizures with or without secondary generalisation and adjunctive therapy for myoclonic and GTC seizures (BNFC).</td>
</tr>
<tr>
<td></td>
<td>Topiramate</td>
<td>At the time of publication, topiramate did not have UK marketing authorisation for use in myoclonic seizures. It had authorisation for monotherapy and adjunctive treatment of focal seizures and GTC seizures and as adjunctive treatment for seizures associated with Lennox–Gastaut syndrome (BNFC).</td>
</tr>
<tr>
<td></td>
<td>Zonisamide</td>
<td>At the time of publication, zonisamide did not have UK marketing authorisation for use in children younger than 18 years of age owing to insufficient data on safety and efficacy (SPC).</td>
</tr>
<tr>
<td>Tonic-atonic seizures</td>
<td>Lamotrigine</td>
<td>At the time of publication, lamotrigine did not have UK marketing authorisation for use in tonic-atonic seizures. It had authorisation for monotherapy and adjunctive treatment of focal seizures, GTC seizures and</td>
</tr>
<tr>
<td>Condition</td>
<td>Medicine</td>
<td>Details</td>
</tr>
<tr>
<td>------------------------------------------------</td>
<td>------------------------</td>
<td>-----------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Partial-onset seizures associated with Lennox–Gastaut syndrome (BNFC).</td>
<td>Topiramate</td>
<td>At the time of publication, topiramate did not have UK marketing authorisation for use in tonic-atonic seizures. It had authorisation for monotherapy and adjunctive treatment of focal seizures, GTC seizures and adjunctive treatment for seizures associated with Lennox–Gastaut syndrome (BNFC).</td>
</tr>
<tr>
<td>Infantile spasms</td>
<td>ACTH (tetracosactide)</td>
<td>At the time of publication, ACTH (tetracosactide) did not have UK marketing authorisation for infantile spasms. Depot ampoules are not recommended in infants and children younger than 3 years owing to the presence of benzyl alcohol in the formulation (SPC).</td>
</tr>
<tr>
<td>Lennox–Gastaut syndrome</td>
<td>Felbamate</td>
<td>At the time of publication, felbamate did not have UK marketing authorisation. There was no SPC available.</td>
</tr>
<tr>
<td>Dravet syndrome</td>
<td>Topiramate</td>
<td>At the time of publication, topiramate did not have UK marketing authorisation for use in Dravet syndrome but did have authorisation for generalised tonic–clonic seizures, focal seizures and seizures associated with Lennox–Gastaut syndrome (BNF).</td>
</tr>
<tr>
<td>BECTS/Panayiotopoulos syndrome and late-onset childhood occipital epilepsy (Gastaut type)</td>
<td>Carbamazepine</td>
<td>At the time of publication, carbamazepine did not have UK marketing authorisation for BECTS/Panayiotopoulos syndrome and late-onset childhood occipital epilepsy (Gastaut type) but had authorisation for focal and generalised tonic–clonic seizures (BNF).</td>
</tr>
<tr>
<td>Lamotrigine</td>
<td></td>
<td>At the time of publication, lamotrigine did not have UK marketing authorisation for BECTS/Panayiotopoulos syndrome and late-onset childhood occipital epilepsy (Gastaut type) but had authorisation for focal and primary and generalised tonic–clonic seizures (BNF).</td>
</tr>
<tr>
<td>Drug</td>
<td>Description</td>
<td></td>
</tr>
<tr>
<td>--------------</td>
<td>---------------------------------------------------------------------------------------------------------------------------------------------</td>
<td></td>
</tr>
<tr>
<td>Oxcarbazepine</td>
<td>At the time of publication, oxcarbazepine did not have UK marketing authorisation for BECTS/Panayiotopoulos syndrome and late-onset childhood occipital epilepsy (Gastaut type) but had authorisation for focal seizures with or without generalised tonic-clonic seizures (BNF).</td>
<td></td>
</tr>
<tr>
<td>Levetiracetam</td>
<td>At the time of publication, levetiracetam did not have UK marketing authorisation for BECTS/Panayiotopoulos syndrome and late-onset childhood occipital epilepsy (Gastaut type) but had authorisation for monotherapy and adjunctive treatment of focal seizures with or without secondary generalisation and adjunctive therapy for myoclonic and GTC seizures (BNFC).</td>
<td></td>
</tr>
<tr>
<td>IGE</td>
<td>Clobazam</td>
<td></td>
</tr>
<tr>
<td></td>
<td>At the time of publication, clobazam did not have UK marketing authorisation for use in children younger than 3 years (BNFC). This was because of insufficient experience of the use of this drug in children younger than 3 years to enable any dosage recommendation to be made (SPC).</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Lamotrigine</td>
<td></td>
</tr>
<tr>
<td></td>
<td>At the time of publication, lamotrigine did not have UK marketing authorisation for use in IGE. It had authorisation for monotherapy and adjunctive treatment of focal seizures, GTC seizures and seizures associated with Lennox–Gastaut syndrome and monotherapy treatment of absence seizures in children (BNF).</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Levetiracetam</td>
<td></td>
</tr>
<tr>
<td></td>
<td>At the time of publication, levetiracetam did not have UK marketing authorisation for IGE but had authorisation for...</td>
<td></td>
</tr>
<tr>
<td>Category</td>
<td>Drug</td>
<td>Information</td>
</tr>
<tr>
<td>--------------------------------</td>
<td>-----------------</td>
<td>----------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Monotherapy and Adjunctive</td>
<td>Topiramate</td>
<td>At the time of publication, topiramate did not have UK marketing authorisation for use in IGE but had authorisation for focal seizures, GTC seizures and seizures associated with Lennox–Gastaut syndrome (BNF).</td>
</tr>
<tr>
<td>Treatment of Focal Seizures</td>
<td>Zonisamide</td>
<td>At the time of publication, zonisamide did not have UK marketing authorisation for use in IGE but had authorisation for adjunctive therapy for adult patients with partial seizures, with or without secondary generalisation (BNF).</td>
</tr>
<tr>
<td>with or without Secondary</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Generalisation and Adjunctive</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Therapy for Myoclonic and GTC</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Seizures</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Juvenile Myoclonic Epilepsy</td>
<td>Clobazam</td>
<td>At the time of publication, clobazam did not have UK marketing authorisation for use in children younger than 3 years (BNFC). This was because of insufficient experience of the use of this drug in children younger than 3 years to enable any dosage recommendation to be made (SPC).</td>
</tr>
<tr>
<td>Lamotrigine</td>
<td></td>
<td>At the time of publication, lamotrigine did not have UK marketing authorisation for use in juvenile myoclonic epilepsy (BNF) but had authorisation for monotherapy and adjunctive treatment of focal seizures, GTC seizures and seizures associated with Lennox–Gastaut syndrome and monotherapy treatment of absence seizures in children (BNF).</td>
</tr>
<tr>
<td>Levetiracetam</td>
<td></td>
<td>At the time of publication, levetiracetam did not have UK marketing authorisation for monotherapy use in JME but had authorisation for monotherapy and adjunctive treatment of focal seizures with or without secondary generalisation and adjunctive therapy for myoclonic and GTC seizures.</td>
</tr>
<tr>
<td>Drug</td>
<td>Information</td>
<td></td>
</tr>
<tr>
<td>--------------</td>
<td>-----------------------------------------------------------------------------</td>
<td></td>
</tr>
<tr>
<td><strong>Topiramate</strong></td>
<td>At the time of publication, topiramate did not have UK marketing authorisation for use in juvenile myoclonic epilepsy (BNF) but had authorisation for focal seizures, GTC seizures and seizures associated with Lennox–Gastaut syndrome (BNF).</td>
<td></td>
</tr>
<tr>
<td><strong>Zonisamide</strong></td>
<td>At the time of publication, zonisamide did not have UK marketing authorisation for use in juvenile myoclonic epilepsy but had authorisation for adjunctive therapy for adult patients with partial seizures, with or without secondary generalisation. (BNF)</td>
<td></td>
</tr>
</tbody>
</table>

### Absence syndromes

**Clobazam**

At the time of publication, clobazam did not have UK marketing authorisation for use in children younger than 3 years (BNFC). This was because of insufficient experience of the use of this drug in children younger than 3 years to enable any dosage recommendation to be made (SPC).

**Lamotrigine**

At the time of publication, lamotrigine had UK marketing authorisation for monotherapy of typical absence seizures for those aged 2–12 years only. There was not authorisation outside of this age range (BNF).

**Levetiracetam**

At the time of publication, levetiracetam did not have UK marketing authorisation for use in absence syndromes but had authorisation for monotherapy and adjunctive treatment of focal seizures with or without secondary generalisation and adjunctive therapy for myoclonic and GTC seizures (BNF).

**Topiramate**

At the time of publication, topiramate did not have UK marketing authorisation for use in absence syndromes but had authorisation for focal seizures, GTC seizures and seizures associated with Lennox–Gastaut syndrome (BNF).
<table>
<thead>
<tr>
<th>Condition</th>
<th>Drug</th>
<th>At the time of publication, please note...</th>
</tr>
</thead>
<tbody>
<tr>
<td>Syndrome</td>
<td>Zonisamide</td>
<td>At the time of publication, zonisamide did not have UK marketing authorisation for use in absence syndromes but had authorisation for adjunctive therapy in the treatment of adult patients with partial seizures, with or without secondary generalisation (BNF).</td>
</tr>
<tr>
<td>Status Epilepticus</td>
<td>Propofol</td>
<td>At the time of publication, propofol did not have UK marketing authorisation for status epilepticus but had authorisation for other conditions. Diprivan 2%, Propofol-Lipuro 2%, and Propoven 2% were not licensed for use in children younger than 3 years; Diprofusor TCI (‘target controlled infusion’) system was not licensed for use in children (BNFC).</td>
</tr>
<tr>
<td></td>
<td>Thiopental sodium</td>
<td>At the time of publication, thiopental sodium did not have UK marketing authorisation for status epilepticus (only if other measures fail, see section 4.8.2 in BNF), by slow intravenous injection (BNF). It is authorised for convulsive states (75 mg to 125 mg or 3 ml to 5 ml of a 2.5% intravenous infusion) (SPC).</td>
</tr>
<tr>
<td></td>
<td>Midazolam</td>
<td>At the time of publication, midazolam buccal liquid and injection did not have UK marketing authorisation for children with status epilepticus (BNF, BNFC).</td>
</tr>
<tr>
<td></td>
<td>Diazepam</td>
<td>At the time of publication, diazepam did not have UK marketing authorisation for Rectubes and Stesolid Rectal Tubes or for use in children younger than 1 year (BNFC).</td>
</tr>
</tbody>
</table>

BECTS, benign epilepsy with centrotemporal spikes; BNF, British national formulary; BNFC, British national formulary for children; GTC, generalised tonic–clonic; SPC, summary of product characteristics.
### Appendix F: Protocols for treating convulsive status epilepticus in adults and children developed by a consensus group under the remit of the British Paediatric Neurology Association (adults published in 2004 and children published in 2011)

**Treating convulsive status epilepticus in adults (published in 2004)**

| General measures |  
|------------------|------------------|
| **1st stage (0–10 minutes)** |  
| - Secure airway and resuscitate | Early status  
| - Administer oxygen |  
| - Assess cardiorespiratory function |  
| - Establish intravenous access |  
| **2nd stage (0–30 minutes)** |  
| - Institute regular monitoring |  
| - Consider the possibility of non-epileptic status |  
| - Emergency AED therapy |  
| - Emergency investigations |  
| - Administer glucose (50 ml of 50% solution) and/or intravenous thiamine (250 mg) as high potency intravenous Pabrinex if any suggestion of alcohol abuse or impaired nutrition |  
| - Treat acidosis if severe |  
| **3rd stage (0–60 minutes)** |  
| - Establish aetiology | Established status  
| - Alert anaesthetist and ITU |  
| - Identify and treat medical complications |  
| - Pressor therapy when appropriate |  
| **4th stage (30–90 minutes)** |  
| - Transfer to intensive care | Refractory status  
| - Establish intensive care and EEG monitoring |  
| - Initiate intracranial pressure monitoring where appropriate |  
| - Initiate long-term, maintenance AED therapy |  
|
Emergency investigations
Blood should be taken for blood gases, glucose, renal and liver function, calcium and magnesium, full blood count (including platelets), blood clotting, AED drug levels; 5 ml of serum and 50 ml of urine samples should be saved for future analysis, including toxicology, especially if the cause of the convulsive status epilepticus is uncertain. Chest radiograph to evaluate possibility of aspiration. Other investigations depend on the clinical circumstances and may include brain imaging, lumbar puncture.

Monitoring
Regular neurological observations and measurements of pulse, blood pressure, temperature. ECG, biochemistry, blood gases, clotting, blood count, drug levels. Patients require the full range of ITU facilities and care should be shared between anaesthetist and neurologist.
EEG monitoring is necessary for refractory status. Consider the possibility of non-epileptic status. In refractory convulsive status epilepticus, the primary end-point is suppression of epileptic activity on the EEG, with a secondary end-point of burst-suppression pattern (that is, short intervals of up to 1 second between bursts of background rhythm).

Emergency AED therapy for convulsive status epilepticus (published in 2004)

<table>
<thead>
<tr>
<th>Premonitory stage (pre-hospital)</th>
<th>Diazepam 10–20 mg given rectally, repeated once 15 minutes later if status continues to threaten, or midazolam 10 mg given buccally. If seizures continue, treat as below.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Early status</td>
<td>Lorazepam (intravenous) 0.1 mg/kg (usually a 4 mg bolus, repeated once after 10–20 minutes; rate not critical). Give usual AED medication if already on treatment. For sustained control or if seizures continue, treat as below.</td>
</tr>
<tr>
<td>Established status</td>
<td>Phenytoin infusion at a dose of 15–18 mg/kg at a rate of 50 mg/minute or fosphenytoin infusion at a dose of 15–20 mg phenytoin equivalents (PE)/kg at a rate of 50–100 mg PE/minute and/or phenobarbital bolus of 10–15 mg/kg at a rate of 100 mg/minute.</td>
</tr>
<tr>
<td>Refractory status**</td>
<td>General anaesthesia, with one of: • propofol (1–2 mg/kg bolus, then 2–10 mg/kg/hour) titrated to effect • midazolam (0.1–0.2 mg/kg bolus, then 0.05–0.5 mg/kg/hour) titrated to effect • thiopental sodium (3–5 mg/kg bolus,</td>
</tr>
</tbody>
</table>
then 3–5 mg/kg/hour) titrated to effect; after 2–3 days infusion rate needs reduction as fat stores are saturated

- anaesthetic continued for 12–24 hours after the last clinical or electrographic seizure, then dose tapered.

\* In the above scheme, the refractory stage (general anaesthesia) is reached 60/90 minutes after the initial therapy.

This scheme is suitable for usual clinical hospital settings. In some situations, general anaesthesia should be initiated earlier and, occasionally, should be delayed.

Experience with long-term administration (hours or days) of the newer anaesthetic drugs is very limited. The modern anaesthetics have, however, important pharmacokinetic advantages over the more traditional barbiturates.

AED therapy must be given in parallel with emergency treatment. The choice of drug depends on previous therapy, the type of epilepsy, and the clinical setting. Any pre-existing AED therapy should be continued at full dose, and any recent reductions reversed.

If phenytoin or phenobarbitale has been used in emergency treatment, maintenance doses can be continued orally or intravenously guided by serum level monitoring. Other maintenance AEDs can be started also, with oral loading doses. Care needs to be taken with nasogastric feeds, which can interfere with the absorption of some AEDs. Once the patient has been free of seizures for 12–24 hours and provided that there are adequate plasma levels of concomitant AEDs, then the anaesthetic should be slowly tapered.
Guidelines for treating convulsive status epilepticus in children (published in 2011)

The original guidelines for the treatment of convulsive status epilepticus (CSE) were published in 2000. They were subsequently adopted by the Advanced Life Support Group (ALSG) and taught in their courses across the UK and Europe. They represent the basis for much of the management of CSE by junior doctors although they are not intended to cover all situations. They are hospital guidelines and take no account of pre-hospital treatment. They do not include infants those born very prematurely and/or less than 28 days of age. Also, they do not cover children who have frequent episodes of CSE for whom an individually tailored guideline is the best option as their seizures may respond better to specific treatments than others.

Generalised convulsive (tonic–clonic) status epilepticus is defined as a generalised convulsion lasting 30 minutes or longer, or repeated tonic–clonic convulsions occurring over a 30 minutes period without recovery of consciousness between each convulsion. However, the guideline stated that ‘for practical purposes, the approach to the child who presents with a tonic–clonic convulsion lasting more than 5 minutes should be the same as the child who is in “established” status – to stop the seizure and to prevent the development of status epilepticus’. The consensus guideline can be seen in the table below.
## Treating convulsive status epilepticus

<table>
<thead>
<tr>
<th>Time</th>
<th>Event Description</th>
<th>Instructions</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 mins</td>
<td>Seizure starts</td>
<td>Check ABC, high flow O₂ if available</td>
</tr>
<tr>
<td>0 mins</td>
<td></td>
<td>Check blood glucose</td>
</tr>
<tr>
<td>5 mins</td>
<td>Midazolam 0.5 mg/kg buccally or Lorazepam 0.1 mg/kg if intravenous access established</td>
<td>Midazolam may be given by parents, carers or ambulance crew in non-hospital setting</td>
</tr>
<tr>
<td>15 mins</td>
<td>Lorazepam 0.1 mg/kg intravenously</td>
<td>This step should be in hospital</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Call for senior help</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Start to prepare phenytoin for 4(^{th}) step</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Re-confirm it is an epileptic seizure</td>
</tr>
<tr>
<td>25 mins</td>
<td>Phenytoin 20 mg/kg by intravenous infusion over 20 mins</td>
<td>Paraldehyde 0.8 ml/kg of mixture may be given after start of phenytoin infusion as directed by senior staff</td>
</tr>
<tr>
<td></td>
<td>or (if on regular phenytoin)</td>
<td>Inform intensive care unit and/or senior anaesthetian</td>
</tr>
<tr>
<td></td>
<td>Phenobarbital 20 mg/kg intravenously over 5 mins</td>
<td></td>
</tr>
<tr>
<td>45 mins</td>
<td>Rapid sequence induction of anaesthesia using thiopental sodium 4 mg/kg intravenously</td>
<td>Transfer to paediatric intensive care unit</td>
</tr>
</tbody>
</table>

When the protocol is initiated it is important to consider what pre-hospital treatment has been received and to modify the protocol accordingly.
Non-convulsive status epilepticus in adults and children

[2004 guideline]

Suggested by the 2004 Guideline Development Group.

This is less common than tonic–clonic status epilepticus. Treatment for non-convulsive status epilepticus is less urgent than for convulsive status epilepticus. Treatment should be considered as follows:

• maintenance or reinstatement of usual oral AED therapy
• use of intravenous benzodiazepines under EEG control, particularly if the diagnosis is not established
• referral for specialist advice and/or EEG monitoring.
Appendix G: Abbreviations and glossary

Abbreviations

- AED anti-epileptic drug
- BECTS benign epilepsy with centrotemporal spikes
- BNF British national formulary
- CSWS continuous spike wave of slow sleep
- CT computed tomography
- ECG electrocardiogram
- EEG electroencephalogram
- ESNs epilepsy specialist nurses
- GTC generalised tonic–clonic
- IGE idiopathic generalised epilepsy
- IUD intrauterine device
- JME juvenile myoclonic epilepsy
- LKS Landau–Kleffner syndrome
- MAE myoclonic-astatic epilepsy
- MRI magnetic resonance imaging
- SPC summary of product characteristics
- SUDEP sudden unexpected death in epilepsy
- VNS vagus nerve stimulation
**Glossary**

**Absence seizure** A seizure characterised by behavioural arrest associated with generalised spike wave activity on EEG.

**Adherence** The extent to which the person’s behaviour matches the prescriber’s recommendations.

**Aetiology** The cause or origin of a disease or disorder as determined by medical diagnosis.

**Anti-epileptic drug (AED)** Medication taken daily to prevent the recurrence of epileptic seizures.

**Attack** An episode in the course of an illness.

**Baseline** The initial set of measurements at the beginning of a study (after run-in period where applicable), with which subsequent results are compared.

**Benign epilepsy with centrotemporal spikes (BECTS)** Benign epilepsy with centrotemporal spikes. An epilepsy syndrome of childhood (5–14 years) characterised by focal motor and/or secondarily generalised seizures, the majority from sleep, in an otherwise normal individual, with centrotemporal spikes seen on EEG.

**Carer** Someone other than a healthcare professional who is involved in caring for a person with a medical condition.

**Childhood absence epilepsy** An epilepsy syndrome with an age of onset 4–9 years, characterised by frequent absence seizures associated with 3 Hz spike wave activity on EEG.

**Clinical presentation** The description of the history and presentation of the clinical condition to the assessing medical team.

**Clinician** A healthcare professional providing direct patient care, for example doctor, nurse or physiotherapist.
Comorbidity Co-existence of more than one disease or an additional disease (other than that being studied or treated) in a person.

Concordance This is a recent term the meaning of which has changed. It was initially applied to the consultation process in which doctor and patient agree therapeutic decisions that incorporate their respective views, but now includes patient support in medicine-taking as well as prescribing communication. Concordance reflects social values but does not address medicine-taking and may not lead to improved adherence.

Continuous spike wave of slow sleep (CSWS) An epilepsy syndrome of onset in children characterised by a plateau and regression of cognitive abilities associated with dramatic increase in spike wave activity in slow wave sleep (> 85% of slow sleep). There may be few seizures at presentation.

Convulsive status epilepticus When a convulsive seizure continues for a prolonged period (longer than 5 minutes), or when convulsive seizures occur one after the other with no recovery between. Convulsive status epilepticus is an emergency and requires immediate medical attention.

Dosage The prescribed amount of a drug to be taken, including the size and timing of the doses.

Dravet syndrome An epilepsy syndrome with onset in infancy, characterised by initial prolonged febrile seizures, subsequent development of multiple seizure types including myoclonic, absence, focal and generalised tonic–clonic seizures, with developmental plateau or regression.

Electrocardiogram (ECG) A test that records the heart’s electrical activity.

Electroencephalogram (EEG) An investigation that involves recording of the electrical activity of the brain. Electrodes are attached to standardised points on the person’s head with collodion. Recordings are usually taken across two points.

Epilepsy A condition in which a person has epileptic seizures.
Epilepsy syndromes Distinctive disorders identifiable on the basis of a typical age of onset, seizure types, specific EEG characteristics, and often other features. Identification of epilepsy syndrome has implications for treatment, management and prognosis.

Epileptic seizure A transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous activity in the brain.

Focal seizures A seizure that originates within networks limited to one hemisphere, discretely localised or more widely distributed. Replaces the terms partial seizures and localisation-related seizures.

Generalised seizure A seizure that originates in, and rapidly engages, bilaterally distributed networks. Such bilateral networks can include cortical and subcortical structures but do not necessarily include the entire cortex (ILAE 2010).

Generalised tonic–clonic (GTC) seizure A seizure of sudden onset involving generalised stiffening and subsequent rhythmic jerking of the limbs, the result of rapid widespread engagement of bilateral cortical and subcortical networks in the brain.

Ictal phenomenology Description or history of ictal events (seizures).

Idiopathic Without known cause.

Idiopathic generalised epilepsy (IGE) A well-defined group of disorders characterised by typical absences, myoclonic jerks and generalised tonic–clonic seizures, alone or in varying combinations in otherwise normal individuals. The EEG is also characteristic, demonstrating a distinct pattern of generalised polyspike wave discharges and/or generalised spike wave on EEG. Of presumed genetic aetiology. The new classification of the ILAE (2010) suggests the terminology should change to genetic generalised epilepsy (GGE).

Indication (specific) The defined use of a technology as licensed by the Medicines and Healthcare products Regulatory Agency (MHRA).
Infantile spasms A specific seizure type presenting in the first year of life, most commonly between 3 and 9 months. Spasms are brief axial movements lasting 0.2–2 seconds, most commonly flexor in nature, involving flexion of the trunk with extension of the upper and lower limbs. They are occasionally referred to as 'salaam seizures'.

Intervention Healthcare action intended to benefit the patient, for example, drug treatment, surgical procedure or psychological therapy.

Juvenile absence epilepsy An epilepsy syndrome with an age of onset of 9–13 years characterised by absence seizures, associated with 3–4 Hz spike wave on EEG. Generalised tonic–clonic seizures may occur.

Juvenile myoclonic epilepsy (JME) An epilepsy syndrome with an age of onset 5–20+ years (peak 10–16 years) characterised by myoclonic seizures that most commonly occur soon after waking. Absence and generalised tonic–clonic seizures may occur in between 50% and 80% of people with JME. EEG demonstrates 3–6 Hz generalised polyspike and wave activity, with photosensitivity in more than 30% of people.

Ketogenic diet A specific diet that is high in fats and low in carbohydrates and protein.

Landau–Kleffner syndrome A very rare epilepsy syndrome with an age of onset of 3–6 years characterised by loss of language (after a period of normal language development) associated with an epilepsy of centrotemporal origin, more specifically bitemporal spikes on EEG with enhancement in sleep or CSWS.

Late-onset childhood occipital epilepsy (Gastaut type) Epilepsy with an age of onset in mid-childhood to adolescence with frequent brief seizures characterised by initial visual hallucinations, ictal blindness, vomiting and post-ictal headache. EEG typically shows interictal occipital spikes attenuated by eye opening.
Lennox–Gastaut syndrome An epilepsy syndrome with an age of onset of 3–10 years characterised by multiple seizure types (including atonic, tonic, tonic–clonic and atypical absence seizures), cognitive impairment and specific EEG features of diffuse slow spike and wave (< 2 Hz).

Monotherapy Treatment with one drug.

Myoclonic-astatic epilepsy (MAE) Also known as Doose syndrome. An epilepsy syndrome with an age of onset of 18–60 months, characterised by different seizure types with myoclonic and myoclonic-astatic seizures seen in all, causing children to fall. The EEG shows generalised spike/polyspike and wave activity at 2–6 Hz.

Myoclonic seizures Sudden ‘brief’ (< 100 ms) and almost shock-like involuntary single or multiple jerks due to abnormal excessive or synchronous neuronal activity and associated with polyspikes on EEG.

Neurological deficit A deficiency or impairment of the nervous system.

Non-convulsive status epilepticus A change in mental status or behaviour from baseline, associated with continuous seizure activity on EEG, which is also seen to be a change from baseline.

Non-epileptic attack disorder (NEAD) A disorder characterised by episodes of change in behaviour or movement, not caused by a primary change in electrical activity of the brain. Movements are varied, and the attacks can be difficult to differentiate from epileptic seizures. Refer to appendix A of the full guideline for the differentiation of epileptic attacks from NEAD and its subgroups.

Older people People over the age of 65 years.

Panayiotopoulos syndrome Epilepsy syndrome presenting in early childhood (mean 4.7 years) with rare seizures that are prolonged. Characterised by autonomic features including vomiting, pallor and sweating, followed by tonic eye deviation, impairment of consciousness with possible
evolution into secondary generalisation. Prognosis is excellent and treatment often unnecessary.

**Pharmacokinetic interaction** The way in which a drug is processed by the body, influencing absorption, metabolism, distribution and excretion.

**Prognosis** A probable course or outcome of a disease. Prognostic factors are patient or disease characteristics that influence the course of a disease. Good prognosis is associated with low rate of undesirable outcomes; poor prognosis is associated with a high rate of undesirable outcomes.

**Provocation** Methods used to provoke seizures, such as hyperventilation, photic stimulation, sleep deprivation and withdrawal of medication.

**Quality of life** A combination of a person’s physical, mental and social wellbeing; not just the absence of disease.

**Refractory status epilepticus** Continued status epilepticus despite treatment with two anticonvulsants in appropriate doses. This can occur in both convulsive and non-convulsive status epilepticus.

**Secondary generalisation seizure** Now referred to as a ‘focal seizure evolving to a bilateral convulsive seizure’.

**Serial seizure** Defined as three or more tonic–clonic seizures in an hour.

**Simple and complex partial epileptic seizures** these terms are no longer recommended. They have been generally replaced with the single word, ‘focal’. Focal seizures should include a clear description of the impairment of consciousness. Should be described when appropriate for seizures, but not to classify specific epilepsy syndromes.

**Spasm** An involuntary sudden onset that may or may not be caused by an epileptic seizure.

**Specialist (as used in this guideline)** For adults: a medical practitioner with training and expertise in epilepsy. For children: a paediatrician with training and expertise in epilepsy.
Status epilepticus (convulsive) See convulsive status epilepticus above.

Sudden unexpected death in epilepsy (SUDEP) Sudden, unexplained, witnessed or unwitnessed, non-traumatic and non-drowning death in people with epilepsy, with or without evidence for a seizure, and excluding documented status epilepticus, in which post-mortem examination does not reveal a toxicological or anatomic cause for death.

Syncope (vasovagal syncopal attack) A brief lapse in consciousness caused by transient cerebral hypoxia. May be caused by many different factors, including emotional stress, vagal stimulation, vascular pooling in the legs, diaphoresis, or sudden change in environmental temperature or body position.

Tertiary centre Specialist care unit. Centre for access to secondary care.

Tonic–clonic seizure An epileptic seizure characterised by initial generalised muscle stiffening, followed by rhythmical jerking of the limbs, usually lasting a few minutes. The person may bite their tongue and may be incontinent. They may feel confused or sleepy afterwards, and take a while to recover fully.

Tonic seizures An epileptic seizure characterised by abrupt generalised muscle stiffening possibly causing a fall. The seizure usually lasts less than a minute and recovery is rapid.

 Unless otherwise stated, taken from Mosby’s Medical, Nursing and Allied Health Dictionary 5th edition and supplemented by the text of the full guideline (2004 Guideline).

1 National Collaborating Centre for Primary Care (2009) Medicines adherence: involving patients in decisions about prescribed medicines and supporting adherence. London: Royal College of General Practitioners

3 Nashef L (1997) Sudden unexpected death in epilepsy: terminology and definitions. Epilepsia 38: S20–S22
### Appendix H: Recommendations to be deleted

<table>
<thead>
<tr>
<th>Recommendation</th>
<th>Replaced by</th>
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| 1.8.13A The newer AEDs gabapentin, lamotrigine, levetiracetam, oxcarbazepine, tiagabine, topiramate and vigabatrin, within their licensed indications, are recommended for the management of epilepsy in people who have not benefited from treatment with the older antiepileptic drugs such as carbamazepine or sodium valproate, or for whom the older antiepileptic drugs are unsuitable because:  
- there are contraindications to the drugs  
- they could interact with other drugs the person is taking (notably oral contraceptives)  
- they are already known to be poorly tolerated by the individual  
- the person is a woman of childbearing potential. | This recommendation has been replaced with new recommendations listed from sections 1.9.3 to 1.9.16 |
| 1.8.13C The newer AEDs gabapentin, lamotrigine, oxcarbazepine, tiagabine, topiramate, and vigabatrin (as an adjunctive therapy for partial seizures), within their licensed indications, are recommended for the management of epilepsy in children who have not benefited from treatment with the older antiepileptic drugs such as carbamazepine or sodium valproate, or for whom the older antiepileptic drugs are unsuitable because:  
- there are contraindications to the drugs  
- they could interact with other drugs the person is taking (notably oral contraceptives)  
- they are already known to be poorly tolerated by the individual  
- the person is a woman of childbearing potential. | This recommendation has been replaced with new recommendations listed from sections 1.9.3 to 1.9.16 |
drugs such as carbamazepine or sodium valproate, or for whom the older antiepileptic drugs are unsuitable because:

- there are contraindications to the drugs
- they could interact with other drugs the child is taking (notably oral contraceptives)
- they are already known to be poorly tolerated by the child
- the child is currently of childbearing potential or is likely to need treatment into her childbearing years.

1.8.14 C Vigabatrin is recommended as a first-line therapy for the management of infantile spasms.

1.9.2 Offer a steroid (prednisolone or tetracosactide*) or vigabatrin as first-line treatment to infants with infantile spasms that are not due to tuberous sclerosis. [new 2011]

1.9.3 Offer vigabatrin as first-line treatment to infants with infantile spasms due to tuberous sclerosis. If vigabatrin is ineffective, offer a steroid (prednisolone or tetracosactide*). [new 2011]

1.8.48A The ketogenic diet should not be recommended for adults with epilepsy.

1.12.2 There is currently no good evidence to support the use of the ketogenic diet in adults with epilepsy. [new 2011]

* Please see appendix E for licensing details.
| 1.8.48C | The ketogenic diet may be considered as an adjunctive treatment in children with drug-resistant epilepsy. |
| 1.12.1 | Refer children and young people with epilepsy whose seizures have not responded to appropriate AEDs to a tertiary paediatric epilepsy specialist for consideration of the use of a ketogenic diet. [new 2011] |
| 1.9.1 | An individual who has prolonged convulsive seizures (lasting 5 minutes or more) or serial seizures (three or more seizures in an hour) in the community should receive urgent care and treatment. |
| 1.14.1.1 | Give immediate emergency care and treatment to children, young people and adults who have prolonged (lasting 5 minutes or more) or repeated (three or more in an hour) convulsive seizures in the community. [2011] |
| 1.9.2 | Rectal diazepam is safe and effective in first-line treatment of prolonged seizures and is recommended in the majority of cases. |
| 1.14.1.2 | Administer buccal midazolam* as first-line treatment in children, young people and adults with prolonged or repeated seizures. Administer rectal diazepam* if buccal midazolam* is not available. If intravenous access is already established and resuscitation facilities are available, administer intravenous lorazepam. [new 2011] |
| 1.9.3 | For many individuals and in many circumstances, buccal midazolam* is more acceptable than rectal diazepam and is easier to administer. It should be used according to an agreed protocol drawn up by the specialist and only used following training. |

* Please see appendix E for licensing details.
1.9.7 Depending on response and the individual's situation, emergency services should be contacted, particularly if:

- seizures develop into status epilepticus
- there is a high risk of recurrence
- this is the first episode
- there may be difficulties monitoring the individual’s condition.

1.14.1.7 Depending on response to treatment, the person’s situation and any personalised care plan, call an ambulance, particularly if:

- the seizure is continuing 5 minutes after the emergency medication has been administered
- the person has a history of frequent episodes of serial seizures or has convulsive status epilepticus, or this is the first episode requiring emergency treatment
- there are concerns or difficulties monitoring the person’s airway, breathing, circulation or other vital signs. [new 2011]

1.10.1 In hospital, individuals with generalised tonic–clonic status epilepticus should be managed immediately, as follows (with local protocols being in place – see suggested guideline in Appendix C¹ of the full guideline):

- secure airway
- give oxygen
- assess cardiac and respiratory function
- secure intravenous (IV) access in a large vein.

1.14.2.1 For children, young people and adults with ongoing generalised tonic–clonic seizures (convulsive status epilepticus) who are in hospital, immediately:

- secure airway
- give high-concentration oxygen
- assess cardiac and respiratory function
- check blood glucose levels using a finger prick test
- secure intravenous access in a large vein.
<table>
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<tr>
<th><strong>1.10.2</strong> Lorazepam should be used as a first-line treatment in status epilepticus (see Appendix C of the full guideline).</th>
<th><strong>1.14.2.2</strong> Use intravenous lorazepam as first-line treatment in hospital in children, young people and adults with ongoing generalised tonic–clonic seizures (convulsive status epilepticus). Use buccal midazolam* if unable to secure immediate intravenous access. Use a maximum of two doses of the first-line treatment (including pre-hospital treatment). See also the suggested protocols in appendix F. [new 2011]</th>
</tr>
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<tbody>
<tr>
<td><strong>1.11.4A</strong> In women of childbearing potential, the risk of the drugs (see 1.8.13A) causing harm to an unborn child should be discussed and an assessment made as to the risks and benefits of treatment with individual drugs. There are currently few data on which to base a definitive assessment of the risks to the unborn child associated with newer drugs. Specific caution is advised in the use of sodium valproate because of the risk of harm to the unborn child.</td>
<td><strong>1.15.1.4</strong> Discuss with women and girls of childbearing potential (including young girls who are likely to need treatment into their childbearing years), and their parents and/or carers if appropriate, the risk of AEDs causing malformations and possible neurodevelopmental delay to an unborn child. Assess the risks and benefits of treatment with individual drugs. There are limited data on risks to the unborn child associated with newer drugs. Specifically discuss the risk of continued use of sodium valproate.</td>
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</table>

* At the time of publication ([month year]), this drug did not have UK marketing authorisation for this indication and/or population (please see appendix E for specific details about this drug for this indication and population). Informed consent should be obtained and documented in line with normal standards in emergency care.
| 1.11.4C | In girls of childbearing potential, including young girls who are likely to need treatment into their childbearing years, the risk of the drugs (see 1.8.13C) causing harm to an unborn child should be discussed with the child and/or her carer, and an assessment made as to the risks and benefits of treatment with individual drugs. There are currently few data on which to base a definitive assessment of the risks to the unborn child associated with newer drugs. Specific caution is advised in the use of sodium valproate because of the risk of harm to the unborn child. |
| 1.15.1.4 | Discuss with women and girls of childbearing potential (including young girls who are likely to need treatment into their childbearing years), and their parents and/or carers if appropriate, the risk of AEDs causing malformations and possible neurodevelopmental delay to an unborn child. Assess the risks and benefits of treatment with individual drugs. There are limited data on risks to the unborn child associated with newer drugs. Specifically discuss the risk of continued use of sodium valproate to the unborn child, being aware that higher doses of sodium valproate (> 800 mg/day) are associated with a greater risk than lower doses (< 800 mg/day). |
| 1.11.11 | Women taking enzyme-inducing AEDs who choose to use depot injections of progesterone should be informed that a shorter repeat injection interval is recommended (10 weeks instead of |
| Based on the clinical experience of the GDG, it was decided that this 2004 recommendation was no longer valid. |
| 112 weeks). | 1.11.17 In all women with epilepsy, seizure freedom during pregnancy should be sought. | 1.15.3.18 Aim for seizure freedom before conception and during pregnancy (particularly for women and girls with generalised tonic–clonic seizures) but consider the risk of adverse effects of AEDs and use the lowest effective dose of each AED. [new 2011] |
| 1.11.23 Routine monitoring of AED levels in pregnancy is not recommended. If seizures increase, or are likely to increase, monitoring of AED levels may be useful to plan or anticipate the extent of change of dose adjustment needed. | 1.15.3.9 Do not routinely monitor AED levels during pregnancy. If seizures increase or are likely to increase, monitoring AED levels (particularly levels of lamotrigine and phenytoin which may be particularly affected in pregnancy) may be useful when making dose adjustments. [new 2011] |
| 1.12.1 People with learning disabilities should receive the same support and care for their epilepsy as the general population. In addition, those with learning disabilities need the care of the learning disabilities team. | 1.16.3.5 Do not discriminate against children, young people and adults with learning difficulties and offer the same investigations and therapies as for the general population. [new 2011] |
| 1.12.2 Learning disabilities are a common association with epilepsy. The management and treatment of the epilepsy should be undertaken by a specialist, working within a | 1.16.3.1 Enable children, young people and adults who have learning disabilities, and their family and/or carers where appropriate, to take an active part in developing a |
| multidisciplinary team. | personalised care plan for treating their epilepsy while taking into account any comorbidities. [new 2011] |

1.16.3.2 Ensure adequate time for consultation to achieve effective management of epilepsy in children, young people and adults with learning disabilities. [new 2011]

1.14.1 The recommendations on choice of treatment and the importance of regular monitoring of effectiveness and tolerability are the same for older people as for the general population.

1.18.1 Do not discriminate against older people and offer the same services, investigations and therapies as for the general population. [new 2011]

1.18.2 Pay particular attention to pharmacokinetic and pharmacodynamic issues with polypharmacy and comorbidity in older people with epilepsy. Consider using lower doses of AEDs and, if using carbamazepine, offer controlled-release carbamazepine preparations. [new 2011]