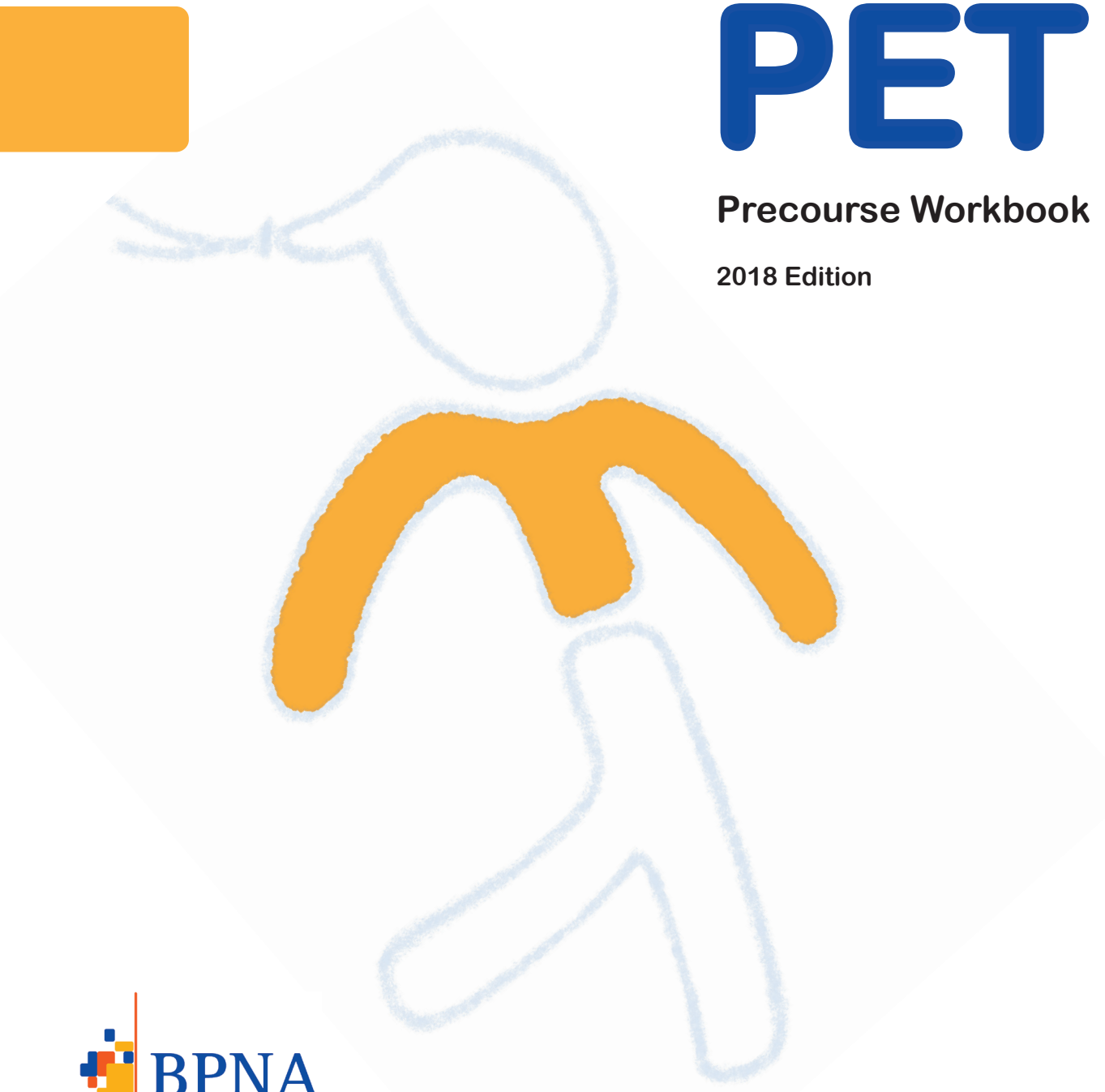
PET 1 2 3
PAEDIATRIC EPILEPSY TRAINING

PET 1

Precourse Workbook

2018 Edition



BPNA
British Paediatric Neurology Association

www.bpna.org.uk

Charity registered in England and Wales (number: 1159115)

Endorsed and supported by
ILAE International League Against Epilepsy

Table of Contents

1. How you should use this handbook

This book is designed to provide you with key facts about childhood epilepsies. It covers the same sort of ground as the training day. It is hoped that by working through the book before the training day you will come to the training day better prepared and therefore more able to fully participate in the sessions.

There are four things you should note:

1. To maintain your interest a number of tasks are given, however, you may not be able to do the tasks. Don't worry! It is hoped that you will be able to after you have completed the PET training.

2. The core material for the course is the text in black and bold. This is considered the essential information, which participants who complete a PET1 course should know at the end of the course. Text highlighted in bold indicates key practice points. Of course, depending on your profession or discipline, these may be more or less important to you.

3. The text in boxes is what the National Institute for Clinical Excellence (NICE), UK says about a particular topic. It is important that you remember that it is not part of the core material and you should not necessarily try to memorise it. However, it is recommended that you at least skim over it. It is included so that if a particular topic is especially relevant to your own practice, you are aware of national policy in the area.

4. Included at the end of each section is additional information about topics covered in the main part of the text. This is material which goes beyond that needed to successfully complete PET1. However, it is hoped that many of you will have your appetite whetted and will wish to know more, perhaps taking a further PET2 or PET3 course. The material here can be seen as a bridge towards these other PET courses. However, if you wish, you can ignore it completely!

Finally, there may be words used in the text with which you are unfamiliar – don't despair. At the end of the book you will find a glossary of terms. Terms which are included in the glossary are indicated in italics.

2. What is and what is not epilepsy?

2.1 Introduction

In this section you will explore what epileptic seizures are, what epilepsy is, and what disorders can be confused with epilepsy.

2.2 Learning objectives

By the end of this section you will:

- Know key epidemiological facts concerning epilepsy
- Know modern terminology used in clinical epileptology
- Be able to give precise definitions of key terms
- Be able to explain in terms understandable to the non-specialist what epileptic seizures are
- Be able to give a simple classification of epileptic seizures and of the different types of epilepsy
- Be able to describe key clinical features which may occur during epileptic seizures
- Be able to list important disorders which may be confused with epilepsy
- Explain why misdiagnosis of epilepsy is common

2.3 Terminology – the good the bad and the ugly!

Task 1:

Having a common vocabulary is essential for effective communication. A bewildering array of terms is used in *epileptology*. Some of these are precise and add clarity when used appropriately, others are imprecise and liable to lead to confusion and some, although still used, are best considered obsolete.

The following is a list of terms that are or have been used in clinical epileptology. Put a tick beside those you think have a precise meaning (and should be used), crosses beside those that you think are obsolete (and best left for the dinosaurs) and question marks besides those that are imprecise and need to be used with care.

Term	✓*Precise*	*X Obsolete*	*? with care*
Convulsion			
Grand mal			
Seizure			
Petit mal			
Epileptic seizure			
Fit			

The only precise term in the list is 'epileptic seizure'. We will look at the definition of this shortly.

The term '*seizure*' can be used to denote any sudden attack from whatever cause. Thus it might be applied to a faint, a severe headache or even a stroke as well as a manifestation of epilepsy. Perhaps its most familiar use, outside epilepsy, is as 'reflex anoxic seizure', a common form of non-epileptic attack mainly occurring in infants and young children. Often in practice the term 'seizure' is often used synonymously with 'epileptic seizure' and this can sometimes be misleading.

The term '*fit*' is used very much in the same way as 'seizure' (i.e. to denote a variety of epileptic and non-epileptic attacks) – it is best avoided.

The term '*convulsion*' is usually used to denote seizures (or fits), in which there is prominent motor activity (such as generalised stiffening, repetitive jerking of the limbs or thrashing movements). Convulsions can be epileptic or non-epileptic.

The terms *grand mal* (literally big attack) and *petit mal* (small attack) were introduced in the 19th century and should no longer be used. They are obsolete principally because what to one person is a grand mal, might be to another a petit mal.

Of the terms indicated **it is best to only use 'seizure' and 'epileptic seizure' and possibly also 'convulsion', but always remembering that neither 'seizure' or 'convulsion' necessarily imply an epileptic basis.**

2.4 Epileptic seizures – what are they?

Task 2:

In Task 1, a distinction was made between *epileptic* and *non-epileptic seizures*. We will now consider what we mean by this distinction. Imagine that on the same day two 12-year-old girls are admitted to the same hospital. Both had been walking to school when they had collapsed to the ground, had gone stiff and had some jerks of their limbs. After a full history had been taken, a medical examination performed and some tests undertaken, one of the girls was diagnosed as having had a probable epileptic seizure, whilst the other was diagnosed as having had a syncopal episode (or 'faint'- an example of a non-epileptic seizure). What is the crucial difference underlying this distinction?

The crucial difference is that **by diagnosing an epileptic seizure one is implying that the attack has occurred as a direct consequence of epileptic activity in the brain** rather than as a consequence of some other mechanism. Note that the outward manifestations of epileptic and non-epileptic seizures may be identical. What is important, in terms of whether they are epileptic or non-epileptic, is the mechanism giving rise to them. Non-epileptic attacks might arise as a consequence of some other disturbance (non-epileptic) of brain activity or else as a consequence of problems outside the brain (for example in the heart).

Of course, by saying that epileptic seizures arise as a consequence of epileptic activity in the brain, one could be accused of simply deflecting the question. What is 'epileptic activity'? An feature of certain brain cells (neurones) is that they are excitable. That is, they can generate and transmit electrical signals. It is disturbances in this that we call epileptic activity. Loosely speaking *epileptic activity* can be considered as a disturbance in the electrical activity of the brain. Stated more scientifically **epileptic activity involves the excessive and/or hypersynchronous discharge of neurones.**

Hence the full definition of an epileptic seizure is:
A transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.

This is the definition given by the International League Against Epilepsy (ILAE). The ILAE is recognised, throughout the world, as the main authority on the use of terminology and classifications in clinical epileptology.

2.5 Epileptic seizures – what happens during them?

Task 3:

The table below lists a whole lot of features that may occur during seizures (epileptic or otherwise). Indicate which you consider might be a manifestation of epileptic activity (i.e. part of an epileptic seizure).		
Feature	*Might be a manifestation of epileptic seizure*	*Not a manifestation of an epileptic seizure*
Sudden fall		
Jerking of limbs		
Blank stare		
Urinary incontinence		
Perceiving a funny smell		
Feeling of fear		
Thrashing movements of limbs		
Facial flushing		
Seeing coloured spots		
Vomiting		
Racing heart		
Tingling sensations		
Headache		
Generalised stiffening		
Floppiness		
Feeling of unfamiliarity		
Ringing noises		
Hiccoughs		
Sudden loss of vision		

Commentary 3:

This was a bit of a trick question. All of the features may be manifestations of epileptic seizures. Remember, **the brain controls the rest of the body, and consequently just about everything imaginable may be a manifestation of epileptic activity**. This may make you think that the task of deciding whether something is likely to be epileptic or not is hopelessly difficult. This is not the case as we shall see later. The point to note at this stage is that there are numerous manifestations of epileptic seizures.

Given the protean manifestations that may occur during epileptic seizures, some order is needed. Hence we classify epileptic seizures into different types. Many different classifications have been devised. **Most modern classifications divide epileptic seizures into *generalised* and *focal epileptic seizures***. The term focal is synonymous but now preferred to *partial*.

Task 4:

Have a stab at trying to define generalised and focal epileptic seizures:	
(i) Generalised epileptic seizure	
(ii) Focal epileptic seizure	

The ILAE gives the following definitions:
Generalised epileptic seizure: ***Are conceptualized as originating at some point within, and rapidly engaging, bilaterally distributed networks. Such bilateral networkscan include cortical and subcortical structures, but not necessarily include the entire cortex.***

Focal epileptic seizure: ***Are conceptualized as originating within networks limited to one hemisphere. These may be discretely localized or more widely distributed***

Put more simply, focal epileptic seizures **start** from a localised area of the brain; generalised epileptic seizures appear to **start** from both sides of the brain simultaneously. The word 'start' is in bold to emphasise that if a seizure starts from a localised part of the brain but then spreads to both sides of the brain, it is still classified as focal. However, in order to indicate the sequence, it can be called a focal seizure evolving to bilateral convulsive seizures (historically referred to as secondary or secondarily generalised seizures).

2.6 Generalised epileptic seizures

There are more than three dozen generalised epileptic seizure types recognised by the ILAE. They are diverse in their manifestations. The most commonly encountered generalised epileptic seizures are the following:

- Generalised tonic clonic seizures (GTCS)
- Tonic seizures
- Myoclonic seizures
- Atonic seizures
- Absence seizures

Task 5:

GTCS constitute what the layman is likely to consider as an epileptic seizure. Write down what you consider to be 3 key features of a GTCS.	
1.	
2.	
3.	

Commentary 5:

GTCS have two main components, the tonic phase and the clonic phase. During the tonic phase the child will go stiff (the meaning of tonic). Simultaneously they may let out a cry and will lose awareness, falling to the ground. After a variable period, the second or clonic phase will begin. It is characterised by rhythmic jerking of the limbs. Note that not all movements during epileptic seizures are clonic – only those involving rhythmical jerking of the limbs. During GTCS many other features may be observed, particularly autonomic features, such as breathing irregularities, colour changes (including cyanosis) and urinary (and occasionally faecal) incontinence. The clonic phase gradually subsides, usually within two minutes or so. Once the seizure stops, the child is likely to be drowsy and often goes to sleep. This is known as the post ictal phase. It may be quite short, lasting a matter of minutes, but can be prolonged for many minutes, or even longer, up to 1-2 hours.

Tonic seizures are characterised by an increase in tone, which may be generalised and obvious or localised and subtle (eg causing retropulsion of the head). *Atonic seizures* involve a loss of postural tone, again this may be generalised and obvious or quite subtle (e.g. causing a head nod). *Myoclonic seizures* (jerks) are characterised by sudden shock like contractions of muscles, or groups of muscles and may be single or repetitive, rhythmical or arrhythmical.

Task 6:

Write down what you think is meant by the term '*absence seizure*'.

In absence seizures the main manifestation is an impairment of awareness. The child may stare blankly ahead and be unresponsive. In some absence seizures other things may happen, for example, the child may fumble with their hands or smack their lips, or the eyelids may blink. However, these features are usually less prominent than the impaired awareness.

There are different types of absences. *Typical absences* start and end abruptly (like a light going off and then coming on again), with the child resuming their normal activities immediately. In *atypical absences* (a different seizure type, which occurs within some epilepsy syndromes, for example *Lennox-Gastaut syndrome*) the start and finish is usually less abrupt, such that the child appears to drift into and drift out of the atypical absence.

In practice, EEG is needed to confirm seizures are absences as there are other types of epileptic and non-epileptic seizures that have 'unresponsive stares' as a feature. The term 'absence' strictly refers to a generalised seizure type with specific EEG changes. The term 'absence' should not be loosely used for any seizure in which impairment of awareness is a feature. For example, focal seizures, especially some arising in the temporal lobes can have altered awareness ore responsiveness as a feature. This change in awareness is sometimes referred to as a dyscognitive feature. For clarity the term absence should be avoided unless specifically referring to an epilepsy with confirmed absence seizures.

2.7 Focal epileptic seizures

The clinical manifestations of focal epileptic seizures depend both on where the seizure starts and where it spreads to.

Until recently focal epileptic seizures were mainly divided into those in which there was impairment of awareness (these were called complex focal or complex partial seizures and are now called dyscognitive) and those in which awareness was retained (these were known as simple focal or simple partial seizures).

Focal seizures are now classified:

(i) With motor or non-motor components (e.g. sensory, autonomic, behaviour arrest, cognitive, emotional)

(ii) With awareness or impaired awareness (previously termed complex partial seizures)

(iii) According to where in the brain they are likely to be arising from. Hence frontal lobe seizures, temporal lobe seizures, parietal lobe seizures and occipital lobe seizures.

2.8 Epilepsy and the epilepsies

So far we have been considering different types of epileptic seizures. Epileptic seizures are merely symptoms, they are not diseases in their own right. A useful analogy is to think about the respiratory system. Cough is an important symptom of respiratory disease. There are different types of cough, such as dry cough, barking cough and productive cough. Although each of these different types of cough tells you something about the patient's condition, they do not define individual diseases. A productive cough can be a symptom of a cold, pneumonia or tuberculosis.

So it is with epileptic seizures. They are symptoms of a whole host of different disorders, which we call epilepsy. Although it is conventional to talk about 'epilepsy', it is important to remember that there is no single entity called epilepsy. Rather the term epilepsy is used to denote a group of heterogeneous disorders in which epileptic seizures occur. **It is better practice to think of 'the epilepsies' rather than 'epilepsy'.**

Task 8:

How might you, in simple terms, define *epilepsy*?

Commentary 8:

The ILAE in 2005 gave the following definition epilepsy:
Epilepsy is a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures, and by the neurobiological, cognitive, psychological, and social consequences of this condition. The definition of epilepsy requires the occurrence of at least one epileptic seizure.

The ILAE in 2014 proposed a new definition:
Epilepsy is a disease of the brain defined by any of the following conditions
1. At least two unprovoked (or reflex) seizures occurring >24 h apart
2.One unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years
3. Diagnosis of an epilepsy syndrome

The important point is that **epilepsy generally involves RECURRENT epileptic seizures.**

Epilepsy is not necessarily life-long. Many children and young people with epilepsy only have epileptic seizures for a matter of months.

The seizures however are not only occurring during some temporary and reversible upset. For example, patients with disturbances of their salt or water balance may have 'epileptic seizures'. However, once the salt or water imbalance is corrected the seizures will stop. These patients are not considered to have epilepsy. Similarly, young children may have 'epileptic seizures' provoked by fever. These are conventionally called febrile seizures and are not considered to indicate epilepsy. Seizures that arise in the context of a temporary, potentially reversible disorders are often called 'acute symptomatic seizures'.

Some different types of epilepsy are called *epilepsy syndromes (sometimes referred to as epilepsy syndromes)*. The ILAE defines an epilepsy syndrome as:

"A complex of signs and symptoms that define a unique epilepsy condition with different aetiologies"

In other words an epilepsy syndrome is a recognisable and characteristic pattern of age of onset, history, examination, seizure type(s) and EEG features. However the underlying cause of the epilepsy for one child with a particular epilepsy syndrome is not necessarily the same as another child with the same epilepsy syndrome.

2.9 Epidemiology

Up to **5% of people will have at least one epileptic seizure in their life.** Of course, not all of these will have recurrent seizures (epilepsy).

- The incidence of epilepsy is the number of new cases diagnosed annually.
- The prevalence of epilepsy is the number of cases of epilepsy at any given time.

Incidence rates vary depending on the definition of epilepsy used and on the age of the population studied. In developed countries the *incidence of epilepsy* is around 150 per 100,000 in the first year of life, 60 per 100,000 in mid-childhood and 45-50 per 100,000 in later childhood.

The *prevalence of epilepsy* in children and young people is 4-5 per 1,000 (0.5% may be easier to remember). For example, in a medium sized UK city like Sunderland, with a total population of 300,000, of which 85,000 are 0-16 years, one would expect there to be about 400 children and young people with epilepsy at any one time.

2.10 What types of epilepsy are there?

The epilepsies are classified according to:

- The types of seizures (generalised or focal)
- Whether they fulfil criteria for a specific epilepsy syndrome
- Any identified underlying cause
- Associated co-morbidities and learning problems

For some children and young people with epilepsy the underlying cause for the epilepsy can be determined. The cause can be a structural, metabolic, infectious, immune and/or a genetic cause. Sometimes the cause is unknown or a combination of the above.

Epilepsy sometimes occurs when an underlying disorder is strongly suspected (e.g. because the child has intellectual disability or severe behavioural problems predating the onset of epileptic seizures), but even after appropriate investigations have been undertaken, no cause can be found. These epilepsies used to be referred to as *probably symptomatic epilepsy* or *'cryptogenic epilepsy.'*

The term Idiopathic epilepsies has fallen in and out of favour within ILAE classification systems. It has been traditionally used for those epilepsies which were presumed to be genetic in origin and are often age-related. The term has created some concern that it encouraged assumptions to be made about a child's diagnosis, intellectual ability or genetic basis that might not be valid. For example, the child had a proven genetic cause for the epilepsy or was unlikely to have intellectual disability. It is currently suggested as a term useful when referring to the following specific epilepsy syndromes: Childhood absence epilepsy, juvenile absence epilepsy, juvenile myoclonic epilepsy, epilepsy with generalised tonic clonic seizures alone.

Task 10:

Write down 5 disorders that you consider may lead to epilepsy as a secondary problem.	
1.	
2.	
3.	
4.	
5.	

Commentary 10:

You could have chosen any of the following:

Brain malformations and maldevelopments

Neurocutaneous disorders (such as tuberous sclerosis)

Post head injury

Post infection (congenital viral infections, meningitis, encephalitis)

Post hypoxic-ischaemic insults (such as birth asphyxia)

Brain tumours

Vascular malformations

Chromosomal abnormalities

Metabolic disorders

2.11 Febrile seizures

By convention epileptic seizures, even if recurrent, which are provoked by and only occur during a temporary disturbance such as a high or low blood sodium level or low blood sugar level are not considered to be a manifestation of epilepsy. It is important to note that, despite this, the seizures are still epileptic in origin. By far the commonest example is febrile seizures.

Most febrile seizures are GTCS. However, fever can provoke other types of epileptic seizures such as generalised clonic seizures, atonic seizures and unilateral seizures (hemiconvulsions). It is also worth remembering that fever can also provoke non-epileptic seizures, such as rigors and faints.

Febrile seizures are conventionally classified as being simple (70%) or complex (30%).

Simple febrile seizures are: generalised (i.e. without focal features, short (last under 10 minutes – some say under 15 minutes) and do not recur within 24 hours, or within the same febrile illness. Complex febrile seizures have focal features, or last more than 10 minutes (some say more than 15 minutes), or recur within 24 hours or during the same febrile illness.

A febrile seizure can be complex because of 1, 2 or 3 of the listed features. Although it is accepted that simple febrile seizures can last 10 (or 15 minutes), in practice, most are much shorter (under 2 minutes). Febrile seizures lasting more than 30 minutes constitute febrile 'status epilepticus' – a medical emergency.

Task 11:

Test your current knowledge of febrile seizures by answering the following questions.	
1.	How young can a child be when they have their first febrile seizure?
2.	To what age can a child continue to have febrile seizures?
3.	How high does the temperature have to be before a febrile seizure can be diagnosed?
4.	Does meningitis cause febrile seizures?
5.	Do febrile seizures run in families?

The National Institute for Health (NIH) in America has defined a febrile seizure as:

"An event occurring in infancy or childhood, usually between 3 months and 5 years of age, associated with fever but without evidence of intracranial infection or defined cause for the seizure"

Other definitions exist but this one is pretty standard. You will note that there is no lower and upper age limits, although to diagnose a febrile seizure under the age of 3 months would be exceptional (and should only be done after the most thorough exclusion of other causes) and very few children will present with a first febrile seizure after the age of 5 years. The peak incidence for the first febrile seizure is from 9 to 20 months.

No definition of fever is given – in practice a temperature of at least 38^{o}C is usually accepted as the lower limit.

By definition, febrile seizures are only diagnosed if there is no other cause for the seizure other than the fever. This means that if a seizure occurs in a child who is febrile as a consequence of meningitis, the seizure is not considered to be febrile. However, it is very important to note that children, particularly young children, with meningitis may have seizures that are indistinguishable from febrile seizures. The definition means that children and young people with other neurological conditions, for example cerebral palsy, who have seizures when febrile should not be diagnosed with febrile seizures.

There is a strong genetic basis for febrile seizures, as indicated by the fact that risk to siblings if one child has had a febrile seizure is about 25% and that there is a high concordance in monozygotic twins. Most authorities consider the inheritance to be polygenic (that is due to the effect of multiple genes), although autosomal dominant inheritance has also been proposed. Recently, linkage to a number of chromosomes has been found in families with febrile seizures, and some children with febrile seizures have been shown to have mutations in specific genes.

Task 12:

You are counselling the mother of a child who has just been diagnosed as having had a febrile seizure. She asks you the following four questions. For each, indicate how you would reply.	
1.	I have never heard of febrile seizure before; are they very rare?
2.	What is the chance that it will happen again?
3.	How dangerous are they?
4.	Does it mean my child will probably develop epilepsy?

By 7 years, 3-4% children will have had 1 or more febrile seizures. Therefore, they are common. Boys are affected more often than girls and black children more often than white children.

The overall risk of recurrence is 30-40%. The main predictors of risk are: early age of onset; family history; duration of illness; and lower temperature at time of seizure. The earlier the age of onset, the greater is the risk of recurrence. Children with a first febrile seizure before one year of age have a 50% chance of recurrence, compared with 20% if the first seizure is after age 3 years. Risk factors can be combined to provide a useful prediction scheme. The recurrence risk for those with none of the four risk factors (age less than 18 months, family history of febrile seizures, low temperature at the time of the seizure and short duration of illness) is 4%, with one factor 23%, with two 32%, with three 62%, and with all four 76%.

Families of children with febrile seizures can be reassured that, with the exception of the risk of injury, short febrile seizures are not dangerous. However, febrile seizures lasting over 30 minutes (febrile status epilepticus) has an appreciable morbidity and mortality. This is largely because febrile status epilepticus may be the presentation of an acute disorder such as meningitis or related to a pre-existing underlying neurological disorder.

Moreover, in most cases febrile seizure will not be followed by epilepsy. However, the risk of the latter is increased compared to the normal population approximately six fold. Risk factors for this are: abnormal neurological or developmental status prior to first febrile seizure (although it is questionable if these should be considered febrile seizures); family history of afebrile seizures; complex febrile seizure. The risk increases the more risk factors there are. For example, if there is a single risk factor, the risk is 6-8% but if all three factors are present it is almost 50%. If epilepsy does develop it can take many different forms.

2.12 Misdiagnosis of epilepsy

The misdiagnosis rate of epilepsy is high. Children and young people are both diagnosed with epilepsy when they do not have epilepsy and, conversely children and young people who have epilepsy are left without the appropriate diagnosis. Recent studies have suggested that **up to a third of children and young people diagnosed with epilepsy may not have it.**

Task 13:

Can you think of some reasons why misdiagnosis of epilepsy is high?

Commentary 13:

Among the more important reasons you could have mentioned:

- In order to make the diagnosis the doctor is usually reliant on descriptions of the attacks; only rarely will he witness them for himself. Such descriptions are likely to be incomplete and inaccurate.
- The clinical events which occur during epileptic seizures often correspond very closely to those which occur during non-epileptic attacks.
- There is no laboratory test for epilepsy, in the way that there is for many other disorders. The best test available, the EEG lacks both sensitivity and specificity. It is liable to misinterpretation. We will look at this in a later section.

2.13 Paroxysmal non-epileptic disorders

There are numerous disorders in which paroxysmal (sudden; unexpected, out-of-the-blue) attacks occur which may mimic or be confused with epileptic seizures. Among the more common and/or important of these are:

- Syncopes and anoxic seizures, including cardiac disorders
  - Reflex anoxic seizures
  - Breath holding attacks
  - Simple faints (vasovagal syncope)
  - Long QT disorders (which predispose to dangerous cardiac arrhythmias)
  - Other cardiac syncopes
  - Suffocation
- Behavioural events and psychological disorders
  - Daydreams and childhood preoccupation / poor ability to concentrate
  - Self-gratification / masturbation
  - Ticks and stereotypies
  - Non-epileptic attack disorder (psychogenic non-epileptic seizures)
- Sleep disorders
  - Nightmares
  - Night-terrors
  - Narcolepsy - cataplexy

- Paroxysmal movement disorders
  - Non-epileptic myoclonus, including benign neonatal sleep myoclonus
  - Dyskinesias
  - Paroxysmal ataxias

During the training day you will have the opportunity to view video examples of many of these and do case studies of common syncopes. For now, it is worth making a few general points:

1. A syncope or faint is a paroxysmal event caused by a sudden, temporary decrease in the supply of oxygenated blood to the brain, either from a reduction in the blood flow itself, or from a drop in the oxygen concentration in the blood, or a combination of both. Syncope is manifested as a loss of awareness, often accompanied by a loss of postural tone sometimes followed by stiffening of the body, jerks, etc. The term anoxic seizure is often used synonymously with syncope, especially if stiffening of the body, jerks, etc. are prominent.

2. Syncopes can be very easily confused with GTCS and some other types of epileptic seizures.

3. There are no single features that reliably distinguish syncopes from epileptic seizures. It is a mistake to rely on features such as the occurrence of urinary incontinence, tongue biting, etc. However, it is usually possible to distinguish between them if a detailed account of all the events that occurred during the attack and the circumstances in which the attack occurred is obtained.

What ILAE says about this

"Epileptic seizures and epilepsy syndromes in individuals should be classified using a multi-axial diagnostic scheme. The axes that should be considered are: description of seizures (ictal phenomenology); seizure types; syndromes and aetiology".

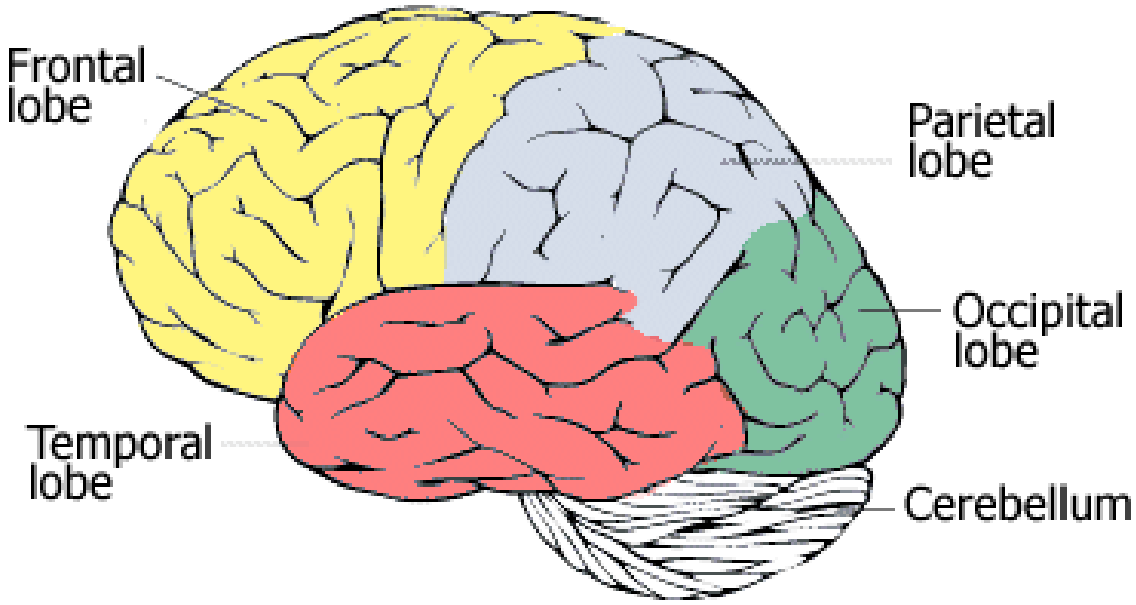
"The seizure types(s) and electroclinical syndrome, aetiology, and co-morbidities should be determined, because failure to classify the electroclinical syndrome correctly can lead to inappropriate treatment and persistence of seizures". BPNA PET courses have developed the DESSCRIBE approach as a pragmatic multiaxial approach. This approach will be introduced and developed throughout the PET courses.

2.14 Additional information

How it possible to work out those parts of the brain which are likely to be involved in a focal epileptic seizure?

Task 14:

The picture of the brain below is annotated to show its component lobes and alongside these are indicated the principle functions of each lobe.



Frontal lobes – major part of the brain controlling movements. Also involved in many higher intellectual functions

Temporal lobes – involved in functions such as emotion and memory. Also involved in the perception of smell and taste and in hearing

Parietal lobes – involved in the perception of 'ordinary' sensations (light touch, pain, etc.)

Occipital lobes – involved in the perception of visual stimuli

The following are brief descriptions of seizures given by, or seen in, children and young people. Indicate, in the table below how the seizures might be classified, by placing a tick in the appropriate box. More than one box may be ticked.

Seizure	*Focal motor seizure*	*Focal sensory seizure*	*Frontal lobe seizure*	*Temporal lobe seizure*	*Parietal lobe seizure*	*Occipital lobe seizure*
1. I start to feel funny. It's horrible and I am afraid. Sometimes I get a horrible taste in my mouth.						
2. It happens at night. He seems to wake up and then have difficulties with his arm and leg. I think the left becomes stiff. Sometimes he makes funny noises. He isn't with it. It's over quickly – after 20 seconds maybe – and then he falls asleep again.						
3. I see blobs of colours, green and red and sometimes purple. They move around a bit but are often at the edge of my vision. I often get a headache. Last time, everything went black – I couldn't see anything. It was very scary.						
4. It starts in my left hand. My fingers twitch. Then my arm starts to go as well and sometimes my face twitches as well.						

Commentary 14:

The first seizure is manifested with fear and unpleasant olfactory hallucinations. These are sensory symptoms (hence it could be classified as a focal sensory seizure) and are often described in patients with temporal lobe seizures.

The second seizure is manifested by motor symptoms involving tonic posturing of limbs. The funny noises are likely to be a manifestation of involvement of the pharyngeal and/or laryngeal muscles. Hence this is a focal motor seizure. It is probably arising in the right frontal lobe.

The third seizure is manifested by visual hallucinations and by blindness. These are sensory symptoms, hence this is a focal sensory seizure. It is likely to arise in the occipital lobes. Headache is common in occipital lobe seizures.

The fourth seizure is manifested as clonic jerking of an upper limb / face. It is clearly a focal motor seizure. It is likely to arise in the primary motor cortex of the right frontal lobe.

3. INVESTIGATIONS

3.1 Introduction

In this section you will explore the uses and abuses of investigations in the diagnosis and management of children and young people with epilepsy or suspected epilepsy.

3.2 Learning objectives

By the end of this section you should:

- Be able to list those investigations which are appropriate when a child newly presents with a seizure.
- Know when it is appropriate to request an EEG.
- Be able to explain the significance of epileptiform and non-epileptiform EEG abnormalities.
- Be able to explain the role of CT and MRI brain scans in investigating children and young people with epileptic seizures.
- Be able to describe when other investigations may be useful.

3.3 The EEG

What is an EEG?

An *EEG* is an investigation in which the electrical activity of the brain is recorded. The activity of neurones generates differences in potential between different parts of the brain. These differences can be detected using electrodes. In most EEG recordings an array of 20 electrodes are used. These are painlessly attached to the scalp usually using paste or a type of glue. This is known as a scalp EEG.

In the past EEGs were recorded onto paper. Now most are recorded digitally and displayed on VDU screens. Most EEGs are done as an out-patient procedure with the patient awake. The EEG is usually recorded for between 20 and 40 minutes. These are called *routine or standard EEGs*. In some departments the routine EEG is combined with a simultaneous video recording. There are a number of special types of EEG. The most common is the *sleep EEG* which as its name suggests is recorded in sleep.

Because epileptic seizures usually only happen occasionally, most EEGs are recorded between seizures (*interictal EEG*). Occasionally, and usually by chance, a seizure occurs while the EEG is being recorded (*ictal EEG*).

What can the EEG do?

Task 1:

Which of the following statements do you think are true?		
		True/False
1.	An abnormal EEG confirms the diagnosis of epilepsy.	
2.	The EEG is a useful test to do if a child's attack is probably non-epileptic but could just possibly be epileptic.	
3.	If an EEG is negative it makes epilepsy unlikely.	

Commentary 1:

You should have answered false to all the questions.

The EEG is an extremely useful test in the investigation of children and young people with epilepsy. However, misuse of it is one of the main reasons why there is a high rate of misdiagnosis.

Any EEG recording consists of the background activity and paroxysmal activity. The former is the on-going electrical activity of the brain. The latter is any burst of EEG activity that stands out as different from the background activity. A normal EEG consists of both normal background activity and normal paroxysmal activity. An EEG can be abnormal either because it contains abnormal background activity or because it contains abnormal paroxysmal activity. However, **only some EEG abnormalities are suggestive of epilepsy**. In particular, some abnormal paroxysmal activity is associated with a much increased risk of epilepsy. Such paroxysmal activity is called *epileptiform activity*. **In general it is only epileptiform activity that supports a diagnosis of epilepsy**. Abnormal background activity and non-epileptiform paroxysmal activity does not, in general, support a diagnosis of epilepsy.

Anyone who requests an EEG should remember 2 key points:

1. **Even in subjects with definite epilepsy a single EEG recording is likely to be normal in about 40-50% of cases (i.e. the EEG lacks sensitivity).**

2. **Abnormal EEGs are common in subjects who do not have and never will have epilepsy. About 5% of healthy children and young people (i.e. who do not have epilepsy) will have epileptiform EEG abnormalities on their EEG (i.e. the EEG lacks specificity).**

It follows from these facts that the EEG cannot be used to either confirm or refute the diagnosis of epilepsy.

Task 2:

We have already considered whether the EEG can help to diagnose epilepsy and that in this regard it has significant limitations. Can you think of any purposes for which the EEG might be useful when investigating a child with epilepsy or suspected epilepsy? [Hint – if you can think of two, you are doing well!]	
1.	
2.	

Commentary 2:

You might have suggested one or more of the following:

(i) Helping to determine the type of epilepsy

(ii) Helping to determine if seizures are precipitated by photic (light) factors

(iii) Helping to decide the child's prognosis

(iv) Helping to decide what drug treatment is the most appropriate

(v) Helping to decide whether to continue or discontinue antiepileptic drug treatment

As we have already seen, there are many different types of epilepsy. Different types of epilepsy are often associated with characteristic epileptiform abnormalities. Hence **the EEG can be very useful in helping decide what type of epilepsy a subject has.** Because this is so useful in determining whether to use antiepileptic drugs to treat a particular patient and, if so, what drug to use, **the EEG can be very helpful in deciding what is the most appropriate drug treatment.** Also, because the best determinant of prognosis is the type of epilepsy, **the EEG can help guide prognosis.**

A small number of children and young people with epilepsy (probably about 5%) have seizures which are precipitated by photic (light) factors, such as TVs, video-games and discos. This *photosensitivity* can be reliably detected using photic stimulation during the EEG recording. **Therefore, the EEG can help decide if a subject's seizures are likely to be provoked by flashing lights, etc.**

It seems common sense that the EEG should be helpful in deciding how long antiepileptic drugs should be continued. Unfortunately, in general, it is far too insensitive for this, but there are some exceptions.

What NICE says about using the EEG

"Individuals requiring an EEG should have the test performed soon after it has been requested" [soon being within 4 weeks]"

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

"An EEG should not be performed in the case of probable syncope because of the possibility of a false-positive result"

"The EEG should not be used to exclude a diagnosis of epilepsy in an individual in whom the clinical presentation supports a diagnosis of a non-epileptic event"

"The EEG should not be used in isolation to make a diagnosis of epilepsy"

"An EEG may be used to help determine seizure type and epilepsy syndrome in individuals in whom epilepsy is suspected. This enables individuals to be given the correct prognosis"

"In individuals presenting with a first unprovoked seizure, unequivocal epileptiform activity shown on EEG can be used to assess the risk of seizure recurrence"

"For individuals in whom epilepsy is suspected, but who present diagnostic difficulties, specialist investigations should be available"

"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"

"Repeated standard EEGs should not be used in preference to sleep or sleep-deprived EEGs"

"When a standard EEG has not contributed to diagnosis or classification, a sleep EEG should be performed"

"In children, a sleep EEG is best achieved through sleep deprivation or the use of melatonin"

"Long-term video or ambulatory EEG may be used in the assessment of individuals who present diagnostic difficulties after clinical assessment and standard EEG"

"Photic stimulation and hyperventilation should remain part of standard EEG assessment. The individual and family and/or carer should be made aware that such activation procedures may induce a seizure and they have a right to refuse"

3.4 Brain scans

CT or MRI?

MRI is the imaging investigation of choice for children and young people with epilepsy. This because of the level of detail MRI shows of the brain structure and it also does not expose the child to ionising radiation as is the case with CT. A CT scan may be indicated in certain situations.

Task 3:

Can you think of one advantage of CT scanning compared to MRI scanning in investigating children and young people with epilepsy and vice versa?

Commentary 3:

In general anything that is likely to be visible on CT scan will be visible on MRI scan and there are many things that are visible on MRI which are not visible on CT scan. (There are two exceptions to this: fresh blood and calcium are usually better seen on CT, unless special MRI sequences are used). This means that **MRI is nearly always preferred over CT when investigating children and young people with epilepsy.**

The major exception to this is when a child presents with a seizure in an emergency, particularly if he or she is not known to have seizures. If he or she recovers as expected, it is not usually necessary to obtain any form of brain scan urgently. However, if the child does not recover as expected, or if there are other worrying features, then a brain scan should be obtained. **In the emergency situation the imaging method of choice is CT** because:

(i) *Intracranial bleeding* is usually an important consideration and is better detected by CT than MRI.

(ii) It is generally much more readily available.

Another consideration, particularly in younger children is that it is often possible to obtain a CT scan without sedation or an anaesthetic whilst this may be needed for an MRI scan. Some children and young people with a developmental age of under 7 years will not lie still for an MRI scan without either sedation or an anaesthetic. However, it should be noted that babies will often lie still if they are fed, wrapped and allowed to fall asleep. It should only be necessary to sedate or anesthetise children and young people for CT scans if they have significant behavioural problems.

Task 4:

Can you think of the sort of things a MRI brain scan might show in?	
a)	A child with an epilepsy where neuroimaging is not typically indicated (for example childhood absence epilepsy, juvenile absence epilepsy, juvenile myoclonic epilepsy)?
b)	A child with epilepsy secondary to a structural cause?

Commentary 4:

(a) This is of course a trick question. By definition **you would not expect there to be any scan abnormalities in these children and young people.**

(b) These epilepsies can be associated with a large number of different abnormalities on MRI scans. These include:

(i) Old destructive lesions following *hypoxic-ischaemic insults*, infections, etc.

(ii) *Malformations* and *maldevelopments* of the brain, such as agenesis of the corpus callosum, generalised neuronal migration defects and focal neuronal migration defects. One can also include here the lesions associated with disorders such as tuberous sclerosis and Sturge Weber syndrome.

(iii) Vascular malformations, such as arteriovenous malformations and cavernous angiomas.

(iv) Brain tumours, particularly slow growing tumours such as gliomas.

(v) *Mesial temporal sclerosis* which is a sclerotic ('scar-type') lesion affecting the medial part of the one or other. *temporal lobes* and is commonly seen in children, young people and adults with temporal lobe seizures.

This list is not exhaustive. In many cases of epilepsy, particularly if long-standing, non-specific abnormalities, such as diffuse *atrophy* will be seen.

Should all children and young people with epilepsy have a scan?

There is a fairly broad consensus as to when scanning is appropriate. This is reflected in the NICE guidelines shown below. Rather than say who should be scanned, it is perhaps easier to state that all children and young people with epilepsy should be scanned unless they are diagnosed as having certain epilepsy syndromes (*juvenile absence epilepsy, juvenile myoclonic epilepsy, childhood absence epilepsy, benign childhood epilepsy with centrotemporal spikes*). *i.e.* those that used to be referred to as 'idiopathic generalised' or 'benign focal' epilepsies.

It should be noted that even if the child is diagnosed with one of these epilepsy syndromes, an MRI scan should be considered if seizures behave uncharacteristically and continue in spite of first-line medication.

What NICE says about this

"Neuroimaging should be used to identify structural abnormalities that cause certain epilepsies"

"MRI should be the imaging investigation of choice in individuals with epilepsy"

"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."

"Individuals requiring MRI should have the test performed soon (within 4 weeks)"

"Neuroimaging should not be routinely requested when a diagnosis of idiopathic generalised epilepsy has been made"

"CT should be used to identify underlying gross pathology if MRI is not available or is contraindicated, and for children in whom a general anaesthetic or sedation would be required for MRI but not CT"

"In an acute situation, CT may be used to determine whether a seizure has been caused by an acute neurological lesion or illness"

3.5 Other tests

Other than an EEG there are no tests which all children and young people with epilepsy must have.

If, when first seen, the child is still convulsing then it is essential to exclude *hypoglycaemia* with a bedside estimation of the blood glucose followed by a laboratory measurement of a true glucose. It is also good practice to exclude *hypocalcaemia* and *hypo/hypernatraemia*. Other investigations will be dictated by the clinical circumstances.

If the child has already stopped convulsing when first seen and is recovering as expected no investigations are necessary.

If a underlying cause for a child's epilepsy is suspected then appropriate *neurometabolic* and *genetic investigations* should be undertaken alongside the MRI. However, these are determined by the other clinical features, not the epilepsy itself. None of them are urgent and, therefore, can be left to the specialist following referral.

One of the important differential diagnoses of epilepsy is *cardiac syncopes*. Features that might suggest a cardiac syncope include:

(i) Attacks manifested by sudden collapses without any warning symptoms (such as usually occur in simple faints).

(ii) Attacks during exercise.

(iii) Family history of *cardiac arrhythmias* and/or sudden death.

In cases where cardiac syncope is suspected, or there is uncertainty about cause, it is important to obtain a 12 lead ECG. SIGN (Scottish Intercollegiate Guidelines Network) goes further and states that all children with a convulsive seizure should have a 12 lead ECG. Syndromes associate with *prolonged QT interval* are particularly important to exclude. Therefore the corrected QT interval should be calculated and compared to controls. If there is still doubt a cardiac referral should be made.

What NICE has to say about this

"In children, other investigations, including blood and urine biochemistry, should be taken at the discretion of the specialist to exclude other diagnoses and to determine an underlying cause of the epilepsy"

"In children, a 12 –lead ECG should be considered in cases of diagnostic uncertainty"

"In cases of diagnostic uncertainty, a referral to a cardiologist should be considered"

3.6 Additional information

What other types of EEG are there?

As was said previously, by far the commonest type of EEG recordings are so-called routine EEGs, but there are a number of other types:

- Sleep deprived and/or Sleep EEG recordings
  These are relatively short (usually about an hour) EEG recordings, during which it is hoped the patient will fall asleep. In order to make sleep more likely the patient may be partially sleep deprived or melatonin may be used to induce sleep. Partial sleep deprivation might involve asking the child's parents to keep them up late the night before the recording and wakening them up early on the day of the recording. Sleep EEGs are usually interictal recordings and are usually done as an out-patient or day case procedure. They are useful because certain EEG abnormalities associated with epilepsy are more common in sleep.

- Ambulatory EEG Recordings
  These are prolonged EEG recordings, usually lasting about 24-hours. The child has the electrodes applied as an out-patient. The data is recorded in a small box. The child is allowed home and carries on normal activities. The next day the electrodes are removed. The purpose of ambulatory EEG recordings is usually to capture one or more seizures (ictal recording). They are only worth doing if seizures are frequent such that there is a fair chance of one occurring during a 24-hour period.

- Video Telemetry
  These are also prolonged EEG recordings, often made over several days or even a week or more. They are usually done as an inpatient procedure. Some epilepsy services are now offering home video telemetry for certain patients in whom this is appropriate. Essentially both an EEG and a simultaneous video are recorded. The purpose is nearly always to record one or more seizures (ictal recording). Video telemetry is an expensive investigation. Its major role is in the evaluation of children and young people for possible surgical treatment of their epilepsy. It is also sometimes used if there is diagnostic doubt as to the nature of a child's attacks.

  All these types of EEG recordings involve applying the electrodes to the scalp (scalp EEG). When children and young people are being

investigated for epilepsy surgery the electrodes are occasionally applied directly onto the brain surface or implanted within the brain substance. These are known as invasive EEG recordings. They require a neurosurgical operation and are only available in specialist epilepsy surgery centres.

If the EEG cannot be used to diagnose epilepsy, is it of no use if I am considering the diagnosis of epilepsy but am not sure?

If, on the basis of the clinical history, an epileptic basis for a child's attacks seems likely, and if the EEG shows epileptiform abnormalities, this can reasonably be said to strengthen the diagnosis. However, if the EEG is normal, the attacks are still quite likely to be epileptic in nature. However, if on the basis of the clinical history a child's attacks are likely to be non-epileptic, the finding of epileptiform EEG abnormalities may be highly misleading. The attacks could still be non-epileptic and the EEG abnormalities could be co-incidental.

Besides CT and MRI scans are there any other types of brain scans used to investigate epilepsy?

In babies whose *fontanelles* are open, ultrasound brain scans can be done. However, these rarely show anything helpful in the diagnosis of epilepsy.

SPECT and PET scans are examples of so-called functional scans. SPECT is usually used to show the pattern of blood flow within the brain, whilst PET is usually used to show the metabolic activity of the brain. These scans are only ever used in the evaluation of children and young people for epilepsy surgery in specialist centres.

4. TREATMENT

4.1 Introduction

In this section you will consider when, why and how we treat children and young people with epilepsy with antiepileptic drugs, including rescue medication. The place of important non-pharmacological treatments will also be looked at.

4.2 Learning objectives

By the end of this section you will:

- Be able to explain the principles that determine whether or not to start antiepileptic drug medication.
- Be able to identify an appropriate antiepileptic drug for treating children and young people with newly presenting seizures.
- Be able to list important adverse effects of antiepileptic drugs.
- Know how to monitor children and young people on antiepileptic drug medication.
- Be able to explain the place of the newer antiepileptic drugs in treating children and young people with seizure disorders.
- Know how and when to discontinue antiepileptic drug medication.
- Understand the role of rescue medication.
- Have an understanding of the role of non-pharmacological treatments for epilepsy

4.3 Why do we treat epileptic seizures?

This seemingly simple question is one of the most important in clinical epileptology.

Task 1:

The following have been suggested as reasons for starting antiepileptic drug treatment in children and young people with epileptic seizures. For each, indicate how important a consideration it is in relation to starting treatment	
a)	To prevent the child suffering unpleasant seizures. *Very important* *Quite important* *Not very important* *Unimportant*
b)	To improve the long term prognosis for seizure control. *Very important* *Quite important* *Not very important* *Unimportant*
c)	To prevent the child dying *Very important* *Quite important* *Not very important* *Unimportant*
d)	To improve the child's performance at school *Very important* *Quite important* *Not very important* *Unimportant*

Commentary 1:

Many epileptic seizures (for example, GTCS and many *temporal lobe seizures*) are unpleasant, frightening and often embarrassing. During them there may be a risk of injury. Antiepileptic drugs will prevent seizures in about 70-80% of cases. **Consequently, the prevention of unpleasant seizures is a major consideration when deciding whether to start antiepileptic drugs.** However, because many subjects will not develop recurrent seizures (i.e. epilepsy) after a single seizure, **there are virtually no circumstances in which antiepileptic drug treatment should be started after a single seizure (status epilepticus is sometimes an exception).**

For many decades most epileptologists considered that 'seizures beget seizures'. In other words that the more seizures one has had the more likely one is to have more. Population studies have shown that in most cases the prognosis for eventual remission is seizures is good and that this does not appear to be determined by whether or not the subject has been treated with antiepileptic drugs. It is generally considered that **about 70-80% of subjects with recurrent epileptic seizures will become seizure free. Consequently, in the vast majority of cases the decision as to whether to start antiepileptic drugs should not be based on the view that not to do so is likely to jeopardise the prospects of long-term seizure control.** An exception to this may be some rare types of epilepsy (the *epileptic encephalopathies*) in which early control of seizures may improve the long-term prognosis – although even this remains to be proven.

The risk of premature death in subjects with epilepsy is 2-3 times higher than that of the general population. People with epilepsy die prematurely for a variety of reasons. Some of these relate to why they have developed seizures. For example, some patients with brain tumours will have seizures and will die prematurely from the tumour and some children and young people with *neurodegenerative diseases* develop seizures and die as a consequence of the neurodegenerative disease. Some people die as a direct result of seizures. For example they may drown in the bath whilst having a seizure, or fall from a height during a seizure. *Status epilepticus* can also directly lead to death.

In addition to these, subjects with epilepsy may die suddenly and unexpectedly. This is known as *SUDEP* (the full definition is that of a sudden, unexpected, witnessed or unwitnessed, non-traumatic and non-drowning death in patients with epilepsy with or without evidence of a seizure, and excluding documented status epilepticus, in which post-mortem examination does not reveal a toxicological or anatomical cause of death). Recently this has been highlighted both in the medical and lay press. SUDEP occurs most often in young adults and the more frequent the seizures the greater the risk.

Nevertheless, despite the increased mortality in epilepsy, the individual risk remains very low and must be balanced against the risks from antiepileptic drugs. **Overall, preventing death is not usually a consideration when deciding whether to treat with antiepileptic drugs.**

Epilepsy can be associated with, in addition to epileptic seizures, cognitive and behavioural problems. However, these are by no means inevitable – many children and young people with quite frequent seizures have few, if any problems at school as a consequence. Nevertheless, there is evidence that in some children and young people, for example, those having absence seizures, antiepileptic drug treatment can improve cognitive function and behaviour and consequently, **improved school performance may be important when considering whether to start antiepileptic drugs.**

To summarise, antiepileptic drugs are usually started after considering fairly obvious issues over the short to medium term, rather than longer term considerations, such as eventual prognosis.

Task 2:

Indicate which management strategy you consider to be the most appropriate in the following situations: [AED = antiepileptic drug treatment]	
a)	A child who has had two febrile seizures. *Regular AED treatment* *No regular AED treatment* *Unsure*
b)	A child who has had 3 GTCS at school. *Regular AED treatment* *No regular AED treatment* *Unsure*
c)	A child who has had 4 nocturnal seizures, characterised by tingling and twitching around the mouth. *Regular AED treatment* *No regular AED treatment* *Unsure*
d)	A 6 year old child who has been diagnosed with frequent typical absence seizures and who is falling behind with her reading. *Regular AED treatment* *No regular AED treatment* *Unsure*

Commentary 2:

(a) The vast majority of children with *febrile seizures* grow out of them without long-term harm. **Treatment of children with febrile seizures with regular antiepileptic drugs is not considered appropriate.**

(b) **GTCS are unpleasant and most authorities would recommend starting treatment after 2 or 3**, especially if they were occurring during the daytime.

(c) There are some childhood epilepsy syndromes which have an excellent prognosis and which are manifested with seizures, which although recurrent, are often not particularly alarming or unpleasant. The most common example of this is *benign childhood epilepsy with centro-temporal spikes (BECTS)* (also called *Rolandic epilepsy*). Seizures in this condition are often fairly 'mild' in their manifestations and usually occur in sleep. The use of antiepileptic drug treatment needs to be carefully weighed up in the individual child, considering the potential impact of seizures and the potential adverse effects of medication.

(d) *Typical absences* in childhood absence epilepsy are 'mild' in their manifestations but usually occur very frequently (sometimes 100s a day). Although major cognitive problems are unusual, minor deterioration in school work is very common and treatment is nearly always considered appropriate.

What NICE has to say on this topic:

"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 individual and their family and/or carers as appropriate"

"AED therapy in children should be initiated by a specialist"

"The decision to initiate AED therapy should be taken between the individual, 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 individual's epilepsy syndrome, prognosis and lifestyle"

"Treatment with AED therapy is generally recommended after a second epileptic seizure"

"AED therapy should be considered and discussed with individuals and their family and/or carers as appropriate after a first unprovoked seizure if:

- The individual has a neurological deficit
- The EEG shows unequivocal epileptic activity
- The individual and/or their family and/or carers consider the risk of having a further seizure unacceptable
- Brain imaging shows a structural abnormality"

"It should be recognised that some individuals (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"

4.4 What drug should I use?

There is rarely a simple answer to this question. Antiepileptic drugs differ in both their spectrums of activity and their adverse affect profiles. It is these that determine which antiepileptic drug is chosen in a given situation. Broadly speaking antiepileptic drugs have either a broad spectrum of activity against both generalised and focal seizure types or a narrow spectrum of activity.

Most children and young people requiring antiepileptic drug treatment are started on either *carbamazepine* or *sodium valproate*. **Carbamazepine has a narrow spectrum of action, mainly being active against focal seizures** (including those evolving to bilateral convulsive seizures). **Sodium valproate has a broad spectrum of action against focal and generalised seizures**. However, there has been mounting concern about the *teratogenic effects* of sodium valproate. Consequently, the committee on the safety of medicines has urged caution in its use in females of child bearing potential. *Lamotrigine or levetiracetam*, newer drugs with a similar spectrum of efficacy to sodium valproate, should be preferred.

What NICE says about this topic:

"The AED treatment strategy should be individualised to the seizure type, epilepsy syndrome, co-medication and co-morbidity, the individual's lifestyle, and the preferences of the individual and their family and/or carers as appropriate"

"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"

4.5 What are the options if initial treatment fails?

The chances of success are good – around 70-80% of children and young people will become seizure free.

Task 3:

List 3 reasons why a child started on antiepileptic drug medication might not become seizure free.	
1.	
2.	
3.	

Commentary 3:

You should have chosen from the following:

- The diagnosis may be wrong – the attacks may not have been epileptic after all – always review the diagnosis.
- The choice of antiepileptic drug may have been wrong for the type of epilepsy – for example, the child may have generalised seizures but has been started on carbamazepine.
- The medication may not have been prescribed appropriately – the dose may have been too low, or the medication may have been given too infrequently (e.g. once rather than twice a day).
- *Concomitant medications* may have been interacting with the drug to reduce its efficacy – always check if the child is taking other medications!
- The child may not be taking the drug as prescribed – poor *compliance.*
- The child may have a *drug resistant epilepsy.*

If having considered other possible reasons for failure, it is concluded that the child's epilepsy is genuinely resistant to the initial antiepileptic drug, there are usually a number of options from which to choose. If the child has focal seizures a drug with a narrow spectrum of action active against focal seizures or with a broad spectrum of action against focal and generalised seizures should be chosen. If the child has generalised seizures a drug with a broad spectrum of activity should be chosen.

4.6 What do I need to think about when using antiepileptic drugs?

The following are the major issues that practitioners need to consider when using antiepileptic drugs.

I. Adverse Effects

Most children and young people have no adverse effects from antiepileptic drugs. However, there are a huge number of potential adverse effects. It is impracticable to memorize these. Far more important is to **warn the parents of children and young people starting antiepileptic drugs always to immediately report anything which causes them concern, particularly**

just after starting medication or following any adjustments. A simple way of classifying the principle adverse effects of antiepileptic drugs is:

(i) *Effects on the CNS* – antiepileptic drugs work in the brain and so it is not at all surprising that all of them can cause CNS side effects. To an extent these effects are predictable, often showing a relationship to the dose used. Problems include headache, drowsiness, irritability, moodiness, hyperactive behaviour and many more besides. Some antiepileptic drugs are more prone to causing CNS adverse effects than others. For example, drowsiness is quite often a problem with carbamazepine, whilst many parents complain that sodium valproate makes their children and young people irritable and moody. Parents often worry that antiepileptic drugs are likely to interfere with a child's learning abilities. However, psychological studies of the common antiepileptic drugs have been very reassuring. Impaired learning abilities are generally only a problem if the drug causes the child to be drowsy.

(ii) *Idiosyncratic reactions* – These are effects which cannot be readily predicted and usually do not show a clear relationship to dose. Many of them are peculiar to particular drugs. Amongst the most common are rashes.

(iii) *Teratogenic effects* – The background risk of a major malformation in the newborn is around 2%. This is increased to 5-6% in those whose mothers received a single antiepileptic drug and to 10% if the mother took two antiepileptic drugs. **All the established antiepileptic drugs have teratogenic effects.** The risk is greatest with sodium valproate. Data is being collected regarding the newer antiepileptic drugs by pregnancy registers. Teratogenic effects for many antiepileptic drugs are lower than for sodium valproate

II. Drug Interactions

Unfortunately most antiepileptic drugs have important interactions to think about. Such interactions include:

(i) Interactions with other antiepileptic drugs. This can lead to increases or decreases in the blood levels of antiepileptic drugs. In turn this can impair the effectiveness of the drug or lead to toxic side effects

(ii) Interactions with other drugs. Some antiepileptic drugs cause the blood levels of other drugs to fall. The most important example of this is that **the efficacy of the contraceptive pill is reduced by a number (but not all) antiepileptic drugs.** Any practitioner starting or

stopping an antiepileptic drug in a woman taking the contraceptive pill needs to consider this carefully. Some drugs can cause an increase or decrease in the levels of anticonvulsant drugs. One important example of this is that **erythromycin (a commonly prescribed antibiotic in children and young people) interacts with carbamazepine, often leading to carbamazepine toxicity.** Drug interactions must be considered when any changes (increase or decrease) are made to the dose of an antiepileptic drug or when any drug is started or stopped in a child on an antiepileptic drug.

III. Blood tests

There is a common misconception that children and young people on antiepileptic drugs require regular blood tests. In fact **most epileptologists manage children and young people with epilepsy with no or only very few blood tests.**

The data sheets of a number of commonly prescribed antiepileptic drugs recommend *full blood counts*, *hepatic* and *renal function tests* prior to and following initiation of the drug. The aim of this is to detect potential adverse effects. However, the evidence that such tests are of practical value is lacking and the NICE guidelines on the management of the epilepsies in children and young people does not recommend them.

It is possible to measure the blood levels of a number of antiepileptic drugs. For some, but not all antiepileptic drugs, there is a correlation between the blood level and effectiveness. However, most children and young people dislike blood tests and in most situations clinical monitoring is at least as effective as blood level monitoring. **Most guidelines do not recommend routine blood level monitoring in children and young people.**

What NICE says about this topic:

"Continuing AED therapy should be planned by the specialist. It should be part of the individual'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"

"The prescriber must ensure that the individual 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"

"Adherence to treatment can be optimised with the following:

- Educating individuals and their families and/or carers in the understanding of their condition and the rationale of treatment
- Reducing the stigma associated with the condition
- Using simple medication regimens
- Positive relationships between healthcare professionals, the individual with epilepsy and their family and/or carers"

"Regular blood test monitoring in children is not recommended as routine, and should only be done if clinically indicated and recommended by the specialist"

"Indications for monitoring 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"

"Asymptomatic minor abnormalities in test results are not necessarily an indication for changes in medication"

"In girls of childbearing potential, including young girls who are likely to need treatment into their childbearing years, the risk of the drugs 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"

"Prescribers should be aware of the latest data on the risks to the unborn child associated with AED therapy when prescribing for women and girls of childbearing potential"

"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"

4.7 When can I stop antiepileptic drug treatment?

In most children epilepsy is not a lifelong condition. In common parlance many children 'grow out of it'. However, this is no means certain and when it occurs is generally unpredictable. **Most children started on antiepileptic drugs will become seizure free and in nearly all there should be an attempt to withdraw antiepileptic medication.** On the basis of outcome studies, it is usually recommended that this be attempted once the child has been free of seizures for 2 years. However, this is not a 'hard and fast rule'. Withdrawal may be considered in some children and young people earlier, in others later and in a very few it may not be appropriate to consider withdrawal. **The single most useful factor in helping to decide when to withdraw antiepileptic drugs is the type of epilepsy or epilepsy syndrome diagnosis.**

What NICE says on this topic:

"The decision to continue or withdraw medication should be taken by the individual, 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 individuals, and their families and/or carers as appropriate, should understand the individual's risk of seizure recurrence on and off treatment. This discussion should take into account details of the individual's epilepsy syndrome, prognosis and lifestyle."

"Withdrawal of AEDs must be managed by, or under the guidance of, the specialist"

"The risks and benefits of continuing or withdrawing AED therapy should be discussed with individuals, and their families and/or carers as appropriate, who have been seizure free for at least 2 years"

"When AED treatment is being discontinued in an individual who has been seizure free, it should be carried out slowly (at least 2-3 months) and one drug should be withdrawn at a time"

"Particular care should be taken when withdrawing benzodiazepines and barbiturate (may take up to 6 months or longer) because of the possibility of drug-related withdrawal symptoms and/or seizure recurrence"

"There should be a failsafe plan agreed with individuals and their families and/or carers as appropriate, whereby if seizures recur, the last dose reduction is reversed and medical advice is sought"

4.8 Rescue medication

The term 'rescue medication' is used to cover the use of antiepileptic drugs acutely to stop seizures rather than regularly to prevent seizures. The management of some children and young people will involve the use of regular antiepileptic drugs and the provision of *rescue medication*. Other children and young people provided with rescue medication will not be treated with regular antiepileptic drugs.

Task 5:

The following statements concern the use of rescue medication. Indicate which you agree with and which you think are incorrect.		
		True/False
a)	All children and young people with epileptic seizures should be provided with rescue medication, because epileptic seizures should be stopped as quickly as possible to minimize brain damage.	
b)	Rescue medication has significant adverse effects and its use should generally be restricted to children and young people with prolonged epileptic seizures.	
c)	There is no place for the use of rescue medication in children and young people with febrile seizures.	

Commentary 5:

You should have answered: False, True, False.

The vast majority of epileptic seizures do not cause harm. Significant concern regarding brain damage is confined to convulsive epileptic seizures lasting longer than 30 minutes. Most convulsive epileptic seizures are short lived (under 2 minutes). **Consequently, most children and young people with seizures do not require rescue medication.**

Although rescue medication is generally safe, significant adverse effects can occur. Of most concern, there is a risk of respiratory depression. On the other hand prolonged convulsive epileptic seizures carry significant risk of permanent brain damage. Since this may occur when a seizure lasts more than 30 minutes, the aim is to stop seizures before the child has been convulsing for 30 minutes. Seizures are usually easier to stop earlier compared to later on. In other words, rescue medication given after 5 minutes is more likely to stop the seizure than if given after 10 minutes. Rescue medication is usually prescribed to children and young people who are considered to be at risk of prolonged epileptic seizures. This is usually on the basis that they have previously had one or more prolonged seizures. The child's carers need to be given explicit instructions as to when the drug should be given. Generally this will be if the child continues to convulse for more than 5 minutes, but depending on the individual, it may be sensible to prescribe a different time. For example, if a child's usual seizures last around 5 minutes, it would be appropriate to give rescue medication for seizures lasting 6 minutes or longer.

One reason for prescribing rescue medication is for children with previous prolonged febrile seizures. Most children with febrile seizures do not require regular antiepileptic drug medication and most febrile seizures are short. However, febrile seizures can continue in some to over 30 minutes (febrile status epilepticus).

There are a number of antiepileptic drugs that can be used as rescue medication. These include:

- Midazolam given *buccally* or *nasally*
- Diazepam given *rectally* (stesolid)
- *Paraldehyde* given rectally

Buccal midazolam is the rescue medication of choice in the UK compared to rectal preparations because of evidence of improved effectiveness and patient preference.

The carers of children and young people prescribed rescue medication must be given detailed training and written instructions on how and when to use it and what to do subsequently. This should include instructions as to when an ambulance should be called. Most ambulance crews will now administer rescue antiepileptic medication to children and young people.

It must be remembered that whilst only a minority of children and young people with epileptic seizures require rescue medication, **the carers of all children and young people with seizures should have an individualised plan as to what to do in the context of a prolonged epileptic seizure or a series of epileptic seizures without full recovery in between seizures.**

What NICE has to say on this:

"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"

"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"

"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"

"Care must be taken to secure the individual's airway and assess his or her respiratory and cardiac function"

Benzodiazepines are the mainstay of first line treatment. "Administer a maximum of two doses of the first-line treatment (including pre-hospital treatment)"

"If seizures continue, administer intravenous phenobarbital or phenytoin as second-line treatment in hospital in children, young people and adults with ongoing generalised tonic–clonic seizures (convulsive status epilepticus)."

"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

4.9 The role of non-antiepileptic drug treatments

There is no evidence to support the use of complementary medicine in the management of children and young people with epileptic seizures. Similarly psychological interventions have not been shown to be effective in the prevention of epileptic seizures, although psychological interventions may be important in managing the co-morbidities associated with epilepsy.

Children and young people whose seizures are resistant to treatment with standard antiepileptic drugs should be referred to a tertiary epilepsy centre. Treatment modalities that are likely to be considered include:

- The use of the newer antiepileptic drugs
- The use of experimental antiepileptic drugs
- Surgical treatment
- The *ketogenic diet*
- *Vagal nerve stimulation*

Surgical treatment of epileptic seizures is possible in a proportion of patients. **Epilepsy surgery has the potential to render some children and young people seizure free (i.e. 'cured') who have hitherto been resistant to antiepileptic drugs. It should be considered in all children and young people with resistant seizures at a reasonably early stage** (generally after 2 drugs have been unsuccessful). Children and young people should be referred to recognised paediatric epilepsy surgery services.

What NICE has to say on this topic:

"If seizures are not controlled and/or there is diagnostic uncertainty or treatment failure, individuals should be referred to tertiary services soon (being seen within 4 weeks) for further assessment. Referral should be considered when one or more of the following criteria are present: …"

"The ketogenic diet may be considered as an adjunctive treatment in children with drug-resistant epilepsy"

"Vagus nerve stimulation is indicated for use as an adjunctive therapy in reducing the frequency of seizures in children who are refractory to antiepileptic medication but who are not suitable for resective surgery. This includes children whose epileptic disorder is dominated by partial seizures (with or without secondary generalisation) or generalised seizures"

4.10 Additional Information

Why did it used to be thought that most people with epilepsy do not recover from it, if in fact, they do?

The main reason for the view that most people with epilepsy would continue to have seizures throughout their lives was because studies were done in big centres. By their very nature these institutions tended to attract the most difficult cases and, therefore, the experience gained in them gave a very warped view.

Some animal experiments also tended to support the notion that recovery from epilepsy was unlikely. Particularly persuasive was the phenomenon of kindling. In this model epileptic seizures are produced using an electrical or chemical stimulus. The more seizures which have been induced the lower the intensity of the stimulus needed to induce further seizures. Eventually spontaneous seizures may occur.

Kindling, although easily induced in rodents is more difficult to induce in higher mammals such as dogs and cats and even more so in primates. Whether it ever occurs in man is unknown. If it does, it is probably uncommon.

People talk about the new and the old antiepileptic drugs. What do they mean by this?

The last two decades has seen a large number of antiepileptic drugs introduced. It has become customary to refer to those available prior to 1989 as the established (or old) antiepileptic drugs and those after 1989 as the new antiepileptic drugs. The established and the new antiepileptic drugs are listed below. Drugs with a narrow spectrum of activity against focal seizures have a suffix (N); those with a broad spectrum of activity have a suffix (B); the spectrums of Phenobarbital and ethosuximide are more complicated

Established antiepileptic drugs

- Phenobarbital (Phenobarbitone)
- Phenytoin (N)
- Carbamazepine (N)
- Ethosuximide
- Benzodiazepines such as clobazam and clonazepam (B)
- Sodium valproate (B)

New Antiepileptic Drugs

- Vigabatrin (N)
- Lamotrigine (N)
- Gabapentin (N)
- Tiagabine (N)
- Oxcarbazepine (N)
- Topiramate (B)
- Levetiracetam (B)
- Zonisamide (N)
- Rufinamide (N)
- Lacosamide (N)
- Perampanel (N)

As previously noted, most children and young people are still started on an established antiepileptic drug as, in general, none of the new drugs have been shown to be clearly superior. However, the concerns regarding the use of sodium valproate in girls and women of potentially child bearing potential has already been noted. In addition, there are two other important exceptions to the rule that most children and young people are started on either carbamazepine or sodium valproate:

- Vigabatrin with or without steroids are particularly effective against epileptic spasms (infantile spasms). It is recommended as first line treatment in these situations and specific treatment choice often depends on whether the child had tuberous sclerosis or not.

- Ethosuximide is a narrow spectrum agent effective against typical absence seizures. It is an alternative to sodium valproate or lamotrigine in childhood absence epilepsy.

If the initial antiepileptic drug fails, how should the second one be introduced?

Two approaches are possible:

- The new drug can be substituted for the first (sequential monotherapy).

- The new drug can be added to the first (polytherapy).

Task 4:

There are advantages and disadvantages of each approach. Can you think of one advantage and one disadvantage of each?
Monotherapy: Advantage - Disadvantage –
Polytherapy: Advantage – Disadvantage –

Commentary 4:

Sequential *monotherapy* has the advantage that it minimises the risk of adverse effects inherent with *polytherapy*. Its disadvantage is that if the drug which is being withdrawn is having some beneficial effect, the patient may experience an increase in seizures. This problem can be reduced if the new drug is introduced whilst the first is being gradually withdrawn.

Polytherapy has the advantage that the patient is not left 'unprotected' during the change-over. However, polytherapy carries with it a significantly greater risk of adverse effects than does monotherapy.

What NICE says about this:

It is recommended that individuals should be treated with a single antiepileptic drug (monotherapy) wherever possible. If the initial treatment is unsuccessful, then monotherapy using another drug can be tried. Caution is needed during the changeover period"

"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"

"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"

"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…."

"The newer AEDs …, 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 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"

"Vigabatrin is recommended as a first-line therapy for the management of infantile spasms"

In what circumstances, if any, is it useful to measure antiepileptic drug levels?

Phenytoin is an antiepileptic drug that is **not** now widely used in children and young people in the UK. For pharmacological reasons, small changes to the phenytoin dose given can lead to very large changes in the levels of drug in the blood. Because of this, children and young people treated with phenytoin require their levels to be checked regularly.

It can also be useful to check levels when making major treatment changes, particularly when using two or more antiepileptic drugs which are known to interact with one another. However, many specialists routinely make such changes without checking levels.

It can be useful to check drug levels if a child is admitted to hospital as an emergency or if poor compliance is suspected.

Although drug levels are usually checked in the blood, they can also be checked in saliva.

5. PSYCHOSOCIAL ISSUES AND EPILEPSY SERVICES

5.1 Introduction

In this section you will take a holistic look at epilepsy, considering how epilepsy impacts on the whole life of the child.

5.2 Learning objectives

By the end of this section you should:

1. Be able to describe the co-morbidities associated with epilepsy.
2. Be able to explain the potential educational impact of epilepsy.
3. Be able to explain why children and young people with epilepsy are stigmatised.
4. Be able to give appropriate safety advise for children and young people with epilepsy.
5. Have knowledge of the social services support available for children and young people with epilepsy and their families.
6. Be able to advise children and young people, parents, carers and other professionals where appropriate information on epilepsy can be obtained.
7. Be able to describe appropriate care pathways for children and young people with epilepsy.

5.3 Epilepsy and co-morbidities

Epilepsy in children and young people is associated with a range of neurodevelopmental, behavioural and psychiatric problems.

Task 1:

Write down three neurodevelopmental problems which you think might be associated with epilepsy.	
1.	
2.	
3.	

Commentary 1:

Children and young people with epilepsy may also have:

- Motor problems, such as *cerebral palsy*
- *Intellectual disability*, which may be mild, moderate or severe
- Behavioural problems
- Sensory problems (particular visual and hearing problems)

These **additional neurodevelopmental problems are, in most cases, not caused by the epilepsy, but rather share the same underlying cause as the epilepsy.** For example, a child who has had birth asphyxia may, as a consequence, develop cerebral palsy, severe intellectual disability, visual problems and epilepsy.

At any particular time one of these problems may be more or less significant.

5.4 Epilepsy and neurodevelopmental problems

Does epilepsy cause neurodevelopmental problems?

This is both a very important question and a very difficult one to answer. Perhaps the best place to start is to remember that epilepsy is not a single disorder, but rather very many different disorders. Some of these .are not usually associated with significant neurodevelopmental problems, others are sometimes associated with neurodevelopmental problems and others are expected to be associated with neurodevelopmental problems.

The current thinking related to evidence arising from research is that the underlying cause of epilepsy (whether identified or not) may impact on neurodevelopment in addition to either seizures or significant electrical discharges, the latter which can be detected by the EEG in the wake or sleep states. Theses frequent discharges are seen in epileptic encephalopathies described below. One may then think of epilepsy in some syndromes as a symptom of an underlying abnormality of brain development.

Some epilepsies are generally not associated with significant neurodevelopmental problems. Examples are juvenile absence epilepsy, juvenile myoclonic epilepsy, childhood absence epilepsy and childhood epilepsy with centrotemporal spikes. Occasionally neurodevelopmental

problems may occur, but these are usually relatively mild. If a child with one of these syndromes presents with developmental or educational problems, referral to a paediatric neurologist is merited.

Another important group of epilepsies are those where an underlying cause is known. **These epilepsies are often associated with pre-existing neurodevelopmental problems.** However, not infrequently (but by no means inevitably) new neurodevelopmental problems develop after the onset of seizures. Children with epilepsy resistant to treatment are more likely to have developmental difficulties.

E*pileptic encephalopathy* is an epilepsy in which cognitive and other impairments have evolved as a direct result of ongoing epileptic activity over and above any underlying diagnosis. Often these epilepsies are resistant to anti-epileptic treatment. Two epilepsy syndromes which you may have heard of which often have an epileptic encephalopathy component are *Lennox Gastaut Syndrome* and *West Syndrome (Epileptic spasms*).

What sort of neurodevelopmental problems can be associated with epilepsy?

When children with epilepsy develop neurodevelopmental problems, these usually take the form of impairments of higher intellectual functions (such as memory impairments, processing speed, visuo-spatial difficulties and language problems) or behavioural problems (such as problems with attention and concentration, hyperactivity and conduct problems). Approximately 40% of school age children with epilepsy show educational underachievement in comparison to their cognitive ability.

Do children with epilepsy often need support at school?

Children with epilepsy often benefit from additional support at school and there is a case for suggesting that every child with epilepsy should have their educational progress monitored more closely than other children. It is important that difficulties are picked up and support provided before children fail as this can affect their confidence and self-esteem and in some children be manifest with behaviour problems at school.

Turning now to the acute effect of seizures. **Temporary brain dysfunction after seizures is common**. This is most dramatically seen following some motor seizures which may be followed by temporary paralysis of a limb or limbs (*Todd's paresis*). Much the same thing can occur following other

seizure types. For example, memory functions may be impaired following some types of seizures. This may be much less obvious to the observer. Although it is temporary, when repeated many times, it may significantly interfere with school progress. In recent years it has been shown that some children and young people have brief impairments of cognitive function during *epileptiform EEG* discharges (not associated with clinical seizures). This is known as *transient cognitive impairment*.

Finally, **antiepileptic drugs may have adverse cognitive and behavioural effects.** Given that antiepileptic drugs act on the brain, it is not surprising that they can cause cognitive and behavioural problems. However, for most antiepileptic drugs used routinely in children and young people such adverse effects are quite rare unless the drug causes drowsiness. Nevertheless, when a child with epilepsy is reported to be having school difficulties, whether in term of learning or behaviour, it is important to consider whether treatment may be causing or contributing to this.

What NICE has to say about this

Neuropsychological assessment should be considered in individuals in whom it is important to evaluate learning disabilities and cognitive dysfunction, particularly in regard to language and memory.
Referral for a neuropsychological assessment is indicated:

- when an individual with epilepsy is having educational or occupational difficulties
- when an MRI has identified abnormalities in cognitively important brain regions
- When an individual complains of memory or other cognitive deficits and/or cognitive decline.

5.5 Epilepsy and psychiatric problems

Psychiatric problems are significantly more common in children and young people with epilepsy than in the general population and are also more common than in children and young people with chronic disorders not involving the CNS. In one study of children and young people with 'uncomplicated' epilepsy:

- 13% had emotional disorder
- 7.5% had conduct disorder
- 5% had mixed psychiatric disorders
- 2% had hyperkinetic disorder

Other psychiatric disorders associated with epilepsy include: autism spectrum disorders and childhood psychoses.

All professionals involved in caring for children and young people with epilepsy should be alert to the occurrence of psychiatric problems, many of which can be helped by suitable and timely interventions. Children and young people with epilepsy should have access to effective psychiatric diagnostic services and treatments.

5.6 Epilepsy and stigma

Stigma occurs when a particular aspect of an individual's behaviour or character is perceived negatively and used to define that person in a negative way.

Task 2:

It should not be difficult to think of reasons why a child with epilepsy's behaviour or character may be perceived negatively. Write a few down:

Commentary 2:

Amongst the many you could have listed are:

- Occurrence of unpredictable, often frightening seizures
- Needs to take drugs
- Reduced school attendance
- Restrictions on activities due to safety considerations
- Presence of co-morbidities (cerebral palsy, intellectual disability, behaviour problems)

In addition to these fairly obvious reasons, false beliefs concerning epilepsy may also contribute to the stigma associated with epilepsy:

- Ideas concerning possession by demons
- Ideas concerning the relationship between epilepsy and sex

5.7 Safety considerations

Most children and young people with epilepsy are restricted in their activities; generally because of fear of death or injury should a seizure occur. **Generally the perceived increased risk of epileptic seizures is much greater than is actually the case**. The general approach should be to encourage normal activities, including sport as much as possible and to minimize risks by taking common sense precautions.

The assessment of risk must be individualised. What might be dangerous for a child having daily drop attacks, might pose no risk for a child with occasional focal seizures occurring without any impairment of awareness.

Generally, the four most risky situations are:

- Water, including swimming and bathing
- Heights
- Heat (cookers, fires, etc.)
- Traffic

There is virtually no evidence that 'ordinary' sports, including contact sports, pose a significant risk, even to those with poorly controlled seizures.

Task 3:

Take a look at the website of SUDEP Action www.epilepsy.org.uk. Read the Epilepsy and Risk – Parent and Carer's Guide (sudep.org/childhood-adolescence-and-risk) Have a look at some of the advice given, for example, about swimming and climbing.

Task 4:

For teenagers, whether or not they will be allowed to drive can be very important. Go again to the website of Epilepsy Action and see what they say about UK driving regulations.

Commentary 4:

It is probably better if you direct patients and their families to suitable advice (web-based or written) concerning psychosocial issues wherever possible as it means they will be able to use these resources to answer other questions which are likely to arise from time to time. Other useful websites are:

Young Epilepsy	www.youngepilepsy.org.uk
National Society for Epilepsy	www.epilepsynse.org.uk
Epilepsy Scotland	www.epilepsyscotland.org.uk

5.8 Organisation of services

The services available for children and young people with epilepsy in the UK are of very variable quality. In 2002 The National Sentinel Clinical Audit of Epilepsy-Related Death was published. The audit considered 22 deaths in children and young people with epilepsy and found that the overall care was inadequate in 77% and that in 59% death may have been possibly or potentially avoidable. Two paragraphs from the report are worth quoting:

"From the available documentation, the audit found deficiencies in access to and quality of care, communication between clinical staff and between healthcare professionals and patients and their carers, documentation and post-mortem investigation of epilepsy-related deaths.

These system failures need to be addressed when planning professional education, clinical and audit guidelines and systems for service delivery. Particular concerns are inadequate access to appropriate epilepsy care; lack of education of healthcare professionals about the principles of epilepsy management and the risks of epilepsy-related deaths; poor communication with patients and their families and between professionals; documentation and post-mortem investigation of epilepsy-related death."

In part the **PET courses** were commenced to address these deficiencies.

Epilepsy12 (www.epilepsy12.com) is a more recent comprehensive UK audit which showed significant improvement but continuing gaps and variation in provision.

There is widespread professional and managerial support for epilepsy care to take place within the context of a managed clinical network. Such a network can be defined as follows:

"Linked groups of health professionals and organisations from primary, secondary, and tertiary care working in a co-ordinated manner, unconstrained by existing professional and organisational boundaries to ensure equitable provision of high quality effective services!"

NICE has given detailed guidance regarding key aspects of service provision. Amongst its key recommendations are:

- First aid advice should be given
- Children and young people with epilepsy should have individualised care plans
- There should be regular specialist review

What NICE says about this:

All individuals with a recent onset suspected seizure should be seen urgently (within 2 weeks) by a specialist (a paediatrician with training and expertise in epilepsy). This is to ensure precise and early diagnosis and initiation of therapy as appropriate to their needs.

NICE gives some more detailed information concerning appropriate action following initial presentation with a seizure:

Individuals 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 when an epileptic seizure is suspected or there is diagnostic doubt.

Protocols should be in place that ensure proper assessment in the emergency setting for individuals presenting with an epileptic seizure (suspected or confirmed).

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

Healthcare professionals should adopt a consulting style that enables the individual 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.

All individuals with epilepsy should have a comprehensive care plan that is agreed between the individuals, their family and/or carers as appropriate, and primary and secondary care providers.

All individuals with epilepsy should have a regular structured review. In children, this review should be carried out at least yearly (but may be between 3 and 12 months by arrangement) by a specialist.

At the review, individuals 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.

NICE has more to say on the provision and role of epilepsy specialist nurses:

Epilepsy specialist nurses (ESNs) should be an integral part of the network of care of individuals 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 individual, families, carers and, in the case of children, others involved in the child's education, welfare and well-being.

If seizures are not controlled and/or there is diagnostic uncertainty or treatment failure, individuals should be referred to tertiary services soon (within 4 weeks) for further assessment.

GLOSSARY

Absence seizure – Strictly speaking an absence seizure is a type of generalised seizure in which the main manifestation is a brief impairment of awareness. However, the term is also sometimes used mistakenly and confusingly to mean any seizure in which the main manifestation is an impairment of awareness.

Anoxic seizure – A term which can be used synonymously with syncope. However, more usually it is reserved for syncopes which include, in addition to a loss of awareness, prominent motor manifestations, such as body stiffening and/or twitching jerking of the limbs

Astatic seizure – An epileptic seizure in which the principle manifestation is a drop to the ground. It is usually caused by a tonic, atonic or myoclonic seizure

Ataxia – A disorder of balance

Atrophy – A term implying shrinkage of a tissue or body part

Atonic epileptic seizure – A type of generalised or focal epileptic seizure manifested by a loss in postural tone which can affect the whole body or only part of it

Atypical absence seizure – A generalised epileptic seizure usually occurring in children and young people with other neurological impairments. Its manifestations are similar to a typical absence seizure but the onset and cessation are often less abrupt and the EEG is different

'Benign epilepsy' – Used to denote an epilepsy which is characterised by epileptic seizures that are easily treated, or require no treatment, and remit without sequelae

Benign childhood epilepsy with centro-temporal spikes - One of the commonest focal epilepsies encountered in otherwise normal school age children

Brain maldevelopment – A term whose meaning is very close to that of brain malformations

Brain malformations – A group of disorders which can give rise to epilepsy in which the brain has developed abnormally in the womb giving rise to a structural abnormality of the brain usually apparent on brain scans, especially MRI brain scans

Breath holding attacks – A disorder, usually encountered in infancy and young children, in which emotional stimuli or minor trauma is quickly followed by non-epileptic syncope. Prior to losing awareness the child often cries and appears to hold the breath in expiration. Breath holding attacks can be divided into **blue** (or **cyanotic**) **breath holding attacks** and **white** (or **pallid**) **breath holding attacks**. The latter is now more correctly termed reflex anoxic seizures

Carbamazepine – A very commonly prescribed antiepileptic drug mainly effective against focal epileptic seizures

Cardiac syncope – A syncope arising as a consequence of dysfunction of the heart. Usually this involves some disturbance of the cardiac rhythm (arrhythmias), but structural heart disease can also give rise to cardiac syncopes

Cataplexy – A rare symptom, often occurring with narcolepsy, in which emotion (such as laughter) triggers a diffuse loss of muscle tone which can mimic an atonic seizure

Cerebral palsy – A term used to denote a group of disorders characterised by abnormalities of movement and posture caused by non-progressive disorders of the developing brain

Chromosomes – The chromosomes are rod shaped structures within the nuclei of cells. They comprise sequences of genes

Compliance – A term denoting adherence to a prescribed drug regime

Concomitant medication – Medication taken at the same time as another medication. In this situation there is always the possibility of drug interactions

Convulsion – The term "convulsion" is a popular, ambiguous, and unofficial term used to mean substantial motor activity during a seizure. In some languages, convulsions and seizures are considered synonyms and the motor component is not clear. (ILAE, 2017). The term in PET courses refers to any seizure in which there is prominent generalised or focal motor activity, such as stiffening, repetitive jerking or thrashing movements. It should not necessarily imply an episode is epileptic or be seen as implying a particular epileptic seizure type.

CT brain scan – A type of scan which produces a series of pictures showing the structure of the brain. It involves the use of x rays

Developmental encephalopathy – developmental impairment without frequent epileptic activity and therefore not associated with regression or further slowing of development (in contrast with *epileptic encephalopathy*)

Diazepam – An antiepileptic drug (also called valium) which is occasionally used as rescue medication and can be given rectally or intravenously

Drop attack – A term synonymous with astatic seizure

Dyskinesias – Non-epileptic disorders associated with abnormal movements

Dyspraxia – A term implying that the subject has problems sequencing together motor tasks. More loosely the term implies extreme clumsiness

ECG – Short for *electrocardiogram.* An investigation, very useful in subjects with suspected disturbances of cardiac rhythm, in which the electrical activity of the heart is recorded using electrodes applied to the chest. The investigation is painless

EEG – Short for *electroencephalogram.* An investigation, very useful in subjects with epilepsy, in which the electrical activity of the brain is sampled using an array of electrodes usually applied to the scalp. The investigation is painless

Encephalitis – Inflammation of the brain, usually caused by virus infections

Epilepsy – A term covering a large group of disorders characterised by the tendency to have recurrent epileptic seizures

Epilepsy surgery – A term used to denote the surgical treatment of epileptic seizures. The surgery either involves removing areas of brain tissue which are giving rise to epileptic seizures (resective epilepsy surgery) or procedures designed to interfere with the spread of epileptic discharges (functional epilepsy surgery)

Epilepsy syndrome – This has historically also been referred to as an 'electroclinical syndrome'. The 'official definition is "A complex of signs and symptoms that define a unique epilepsy condition with different aetiologies". More loosely put an epilepsy syndrome is a type of epilepsy

Epileptic activity - epileptic activity involves the excessive and/or hypersynchronous discharge of neurones (brain cells). Loosely speaking, it can be considered as a disturbance in the electrical activity of the brain

Epileptic encephalopathy - A condition in which epileptic activity in the brain is believed to contribute to a progressive disturbance in cerebral function. More loosely, an epileptic encephalopathy is a type of epilepsy in which continued epileptic seizures or just epileptic discharges in the brain not giving rise to overt seizures, are considered to lead to further problems such as intellectual disability and severe behavioural problems

Epileptic seizure – A type of seizure which arises as a consequence of epileptic activity in the brain. The manifestations of epileptic seizure are protean. The 'official' definition of an epileptic seizure is 'manifestation(s) of epileptic (excessive and/or hypersynchronous), usually self-limited activity of neurones in the brain

Epileptiform activity – Abnormalities seen on the EEG which are strongly associated with an increased risk of epilepsy

Epileptology – The medical specialty concerned with the diagnosis and treatment of the epilepsies

Faint (also called **simple faint** and **vasovagal syncope**) – Overall the commonest form of syncope. Faints can have various triggers, but all are characterised by a temporary reduction in the blood flow to the brain sufficient to cause loss of awareness

Fit – A rather imprecise term usually used synonymously with *epileptic seizure*

Focal motor seizure - A focal epileptic seizure type with prominent motor features, such as jerking or thrashing of the limbs

Focal seizure – Conceptualized as originating within networks limited to one hemisphere. These may be discretely localized or more widely distributed. More loosely stated, a focal epileptic seizure is one which starts from a localized part of the brain

Focal sensory seizure – A focal epileptic seizure with prominent sensory symptoms, such as hallucinations of smell, taste, hearing and vision or 'experiential phenomena' such as feelings of fear, *déjà vu*, etc.

Frontal lobe seizure – A focal epileptic seizure arising from one or other of the frontal lobes of the brain. The frontal lobes are involved in the control of movements and in various higher cognitive functions. The manifestations of frontal lobe seizures reflect these functions

Full blood count – Shorthand for a series of blood tests in which the concentration of haemoglobin and of various cells in the blood is measured

Generalised seizure – *Conceptualized as originating at some point within, and rapidly engaging, bilaterally distributed networks. …can include cortical and subcortical structures, but not necessarily include the entire cortex.* More loosely stated, a generalised epileptic seizure is one which starts from both sides of the brain simultaneously

Generalised tonic clonic seizure – A type of epileptic seizure in which the subject simultaneously loses awareness and becomes stiff all over (the tonic phase). This is then followed by repetitive jerking of all four limbs (the clonic phase).

Genetic investigations – Chromosome and DNA tests usually done on blood samples (although occasionally other tissues such as skin are used) used to detect genetic disorders

Grand mal – An imprecise term (best avoided) used to denote 'major' epileptic seizures. What is meant by 'major' varies from one practitioner to another, but will include GTCS

Hepatic (liver) function tests – Blood tests commonly used to screen for liver disease/dysfunction

Hypernatraemia - A term indicating an abnormally high blood sodium level

Hypocalcaemia - A term indicating an abnormally low blood calcium level

Hypoglycaemia – A term indicating an abnormally low blood sugar level

Hyponatraemia - A term indicating an abnormally low blood sodium level

Hypoxic-ischaemic insult – An insult, for example to the brain, caused by a lack of blood and/or oxygen

Ictal – A term meaning '*seizure*'. An ictal EEG is an EEG during which one or more seizures were recorded

Idiopathic – A term referring to a specific group of epilepsies which comprises childhood absences epilepsy, juvenile absence epilepsy, juvenile absence epilepsy and epilepsy with generalised tonic clonic seizures alone. It's use should not imply any genetic or intellectual features.

Idiopathic generalised epilepsy – A group of epilepsies for which no cause can be found, occurring in otherwise normal subjects and characterised by the occurrence of one or more generalised seizure types. This term was withdrawn from ILAE recommendations in 2011

Idiosyncratic reactions – Unexpected/unpredictable adverse drug effects

Incidence (of epilepsy) – The number of new cases (of epilepsy) diagnosed annually in a given population

Interictal – Between seizures. Hence an interictal EEG is one recorded between seizures

International League Against Epilepsy (ILAE) – In its own words:
The International League Against Epilepsy (ILAE) is the world's pre-eminent association of physicians and other health professionals working towards a world where no persons' life is limited by Epilepsy. Its mission is to provide the highest quality of care and well-being for those afflicted with the condition and other related seizure disorders.

The League aims:

- To advance and disseminate knowledge about epilepsy
- To promote research, education and training
- To improve services and care for patients, especially by prevention, diagnosis and treatment

Intracranial bleeding – Bleeding into or around the brain

Ketogenic diet – A treatment for drug resistant epilepsy which involves giving the patient a diet very high in fats. The excess fats are converted into ketones, the presence of which appears to exert an antiepileptic effect

Lamotrigine – An antiepileptic drug active against both focal and generalised epileptic seizures and epilepsies

Intellectual disability – A term used to denote an incomplete or arrested development of the mind. Moderate intellectual disability (synonymous with mild mental retardation) implies a score on IQ type tests of under 70. Severe intellectual disability (synonymous with severe mental retardation) implies a score on IQ type tests of under 50

Lennox Gastaut syndrome – An epileptic encephalopathy usually occurring in early to mid childhood

Localisation-related epilepsy - Synonymous with 'focal epilepsy'. Focal epilepsy is the preferred term

Localisation-related seizure - Synonymous with 'focal seizure'. Focal seizure is the preferred term

Long QT syndromes – A group of cardiac disorders characterised by the occurrence of syncopes associated with a characteristic appearance on ECG traces. They can be associated with sudden death and are sometimes misdiagnosed as epilepsy

Mesial temporal sclerosis – A common cause of temporal lobe epilepsy. The term implies scarring of the structures lying in the medial part of one or other of the temporal lobes

Metabolic disorders – A large group of generally very rare conditions many of which affect the brain and include in their manifestations epileptic seizures. Their main feature in common is that they involve some problem interfering with the myriad of metabolic pathways in the body. These pathways are responsible, for example, for how the body handles food and stores and utilizes energy

Meningitis – Inflammation of meninges around the brain, usually caused by infections

Mental retardation – see preferred term of '*intellectual disability*'

Midazolam – An antiepileptic drug usually used as rescue medication and usually given buccally (into the cheeks)

MRI brain scan – A type of brain scan which produces a series of pictures showing the structure of the brain. It is generally far more sensitive than CT. It involves the use of very strong magnets

Myoclonic seizure – A type of generalised epileptic seizure characterised by a sudden shock like contraction of a muscle or a group of muscles. NB. Not all types of myoclonus are epileptic

Narcolepsy–cataplexy - a lifelong neurological disorder of state boundary control in which the distinctions between sleep states, particularly REM sleep, and wakening are blurred.

Neurocutaneous disorder – The neurocutaneous disorders are a group of condition which, for embryological reasons, have both skin abnormalities and brain malformations/maldevelopments. They are commonly associated with epilepsy. Examples include tuberous sclerosis, neurofibromatosis and Sturge Weber syndrome

Neurodegenerative disease – A disease in which there is a loss of acquired skills caused by the death of nerve cells. These conditions commonly give rise to dementia. Many metabolic disorders behave in this manner. Epilepsy is common in neurodegenerative diseases

Neurodevelopmental problems – A broad term used to indicate that a child has motor, sensory, cognitive and/or behavioural problems

Neurometabolic investigations – A series of blood, urine and sometimes other tests used to detect metabolic disorders

Non-epileptic seizure – A seizure which is caused by any mechanism other than epileptic activity in the brain. Common types of / causes of non-epileptic seizures include faints, reflex anoxic seizures, cardiac syncopes, psychogenic attacks and some movement disorders

Occipital lobe seizure - A focal epileptic seizure arising from one or other of the occipital lobes of the brain. The occipital lobes are involved in the perception of visual stimuli. The manifestations of occipital lobe seizures reflect this function

Paraldehyde – An antiepileptic drug usually used as rescue medication and given rectally

Parietal lobe seizure – A focal epileptic seizure arising from one or other of the parietal lobes of the brain. The parietal lobes are involved in the perception of sensations such as touch, temperature and pain. The manifestations of parietal lobe seizures reflect this function

Paroxysmal disorders – A term synonymous with *seizure disorder*, when the term *seizure* is taken to include both epileptic and non-epileptic seizures. In other words a paroxysmal disorder is any condition characterised by recurrent epileptic or non-epileptic seizures

Partial seizure – Synonymous with 'focal seizure'. Focal seizure is the preferred term

Photosensitivity – A term implying that a subjects epileptic seizures are likely to be triggered by photic (light) factors. The term is also used to signify the occurrence of certain epileptiform abnormalities on the EEG in response to intermittent photic stimulation

Prevalence (of epilepsy) – The number of cases (of epilepsy) at any given time in a particular population

Probably symptomatic – A term implying that although an underlying cause for the condition cannot be found, such a cause is strongly suspected. Hence a probably symptomatic epilepsy is one for which no cause can be found after appropriate investigations, but because of other problems, such as intellectual disability or behavioural difficulties, an underlying cause responsible for the seizure and the other problems is strongly suspected. This term was withdrawn from ILAE recommendations in 2011

Reflex anoxic seizure – A disorder, mainly of infancy and early childhood characterised by anoxic seizures due to temporary pauses in the heart rhythm and usually triggered by minor bumps. An alternative term is *pallid* or *white breath-holding attacks*

Renal (kidney) function tests – Blood tests used to screen for kidney disease/dysfunction and for abnormalities in the concentrations of salts, such as sodium, in the blood

Rescue medication – When used in the context of epilepsy, this term denotes the use of antiepileptic drugs to stop prolonged epileptic seizures or clusters of epileptic seizures

Resistant (drug resistant) epilepsy – A term implying the continuation of epileptic seizures despite appropriate treatment. Epilepsies are often said to be drug resistant after there has been a failure to respond to two suitable antiepileptic drugs at adequate doses.

Rolandic epilepsy – An alternative name for benign childhood epilepsy with centro-temporal spikes

Seizure – A broad term which can be used to denote any paroxysmal attack. The term applies to, amongst others, attacks such as faints and collapses of cardiac origin as well as to epileptic seizures. However, it is often used interchangeably with *epileptic seizure* which can cause confusion

Semiology – Meaning the clinical features

Sodium valproate – A commonly prescribed broad spectrum antiepileptic drug

Status epilepticus – An epileptic seizure lasting longer than 30 minutes or a series of epileptic seizures over a period of 30 minutes without full recovery between seizures

SUDEP – An acronym for sudden, unexpected, witnessed or unwitnessed, non-traumatic and non-drowning death in patients with epilepsy with or without evidence of a seizure, and excluding documented status epilepticus, in which post-mortem examination does not reveal a toxicological or anatomical cause of death

Syncope – A term used to denote a type of non-epileptic seizure caused by a temporary mismatch in the supply of blood and/or oxygen to the brain in relation to its needs. The cardinal feature of syncopes is loss of (or impairment) in awareness

Symptomatic - A term implying that the underlying cause for the particular condition is known. Therefore, a symptomatic epilepsy is one caused by a known disorder affecting the brain. This term was withdrawn from ILAE recommendations in 2011

Temporal lobe seizure - A focal epileptic seizure arising from one or other of the temporal lobes of the brain. The temporal lobes are involved in the perception of sound, taste and smell and in the experience of emotion. The manifestations of temporal lobe seizures reflect these functions

Teratogenicity – A term indicating the capacity of an agent (e.g. an infection, drug or irradiation) to cause damage to the unborn child

Tonic seizure – A type of generalised epileptic seizure whose principle manifestation is stiffening (which can affect the whole body or only part of it)

Transient cognitive impairment – Brief impairment of cognitive functions associated with epileptiform EEG discharges

Typical absence seizure - A generalised seizure type usually occurring in otherwise healthy children and young people and mainly manifest with a brief impairment of awareness of abrupt onset and cessation. The seizure is accompanied by a characteristic EEG appearance known as '3Hz spike and wave'

Vagal nerve stimulation - A treatment for drug resistant epilepsy which involves implanting an electrical stimulator under the skin of the chest wall. This is used to stimulate the vagus nerve in the neck. This stimulation exerts an antiepileptic effect in the brain

Vascular malformation – An abnormality consisting of abnormally formed blood vessels. Some vascular malformations are located within the brain. They can give rise to various problems, such as bleeding and epilepsy

West syndrome – An epileptic encephalopathy occurring in babies and infants and characterised by a special type of epileptic seizure known as epileptic spasms (previously called infantile spasms)