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The new classification of seizures: an overview for the general physician

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Abstract

The International League Against Epilepsy Classification of the Epilepsies, first presented in 1981, has been widely adopted across the globe. In 2017 it was revised to allow for more robust, specific, flexible and logical classification of seizures. A number of new seizure types are recognised. Classification should be timely as it plays a vital role in the diagnosis and management of patients with epilepsy. Accurate classification also underpins epilepsy research from pathophysiology to public health. Here we review the basic and extended forms of the classification. Semiology (symptoms and signs) is used as the foundation for grouping seizures under focal, generalised or of unknown onset. Focal seizures can be further classified by the presence or absence of awareness and motor signs. Generalised seizures engage bilateral networks from the onset and these can be either motor or non-motor. Seizures of unknown onset can be classified as motor, non-motor, tonic–clonic, epileptic spasms, or behaviour arrest.

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Introduction

Epilepsy is a highly prevalent condition.[1] About 10–20% of acute medical admissions in the UK are neurological;[2,3] of these, a large proportion is due to seizures (see e.g. Nitkunan et al.[4]). There are significant deficiencies in diagnosis and subsequent care revealed by the National Audit of Seizure Management in Hospitals.[5,6] Classifying seizures is very important and depends mainly on their semiology (signs and symptoms), with ancillary investigations being helpful in some cases. Correct classification is critical for practising clinicians in the diagnosis and management of patients with epilepsy.

The International League Against Epilepsy (ILAE) is the most important international organisation dedicated to clinical care, education and research in epilepsy. In 1981 it issued a classification for epilepsy that has been very widely adopted.[7] Recognising a number of deficiencies in this classification, the ILAE released a new seizure classification in 2017.[8,9]

We outline the key points of the basic and expanded seizure classifications. We hope this review will encourage paediatricians and general physicians to adopt the new classification scheme.

Basic seizure classification

For practitioners not having an expertise in epilepsy the basic seizure classification was devised.

Where does the seizure start?

The initial categorisation relates to the onset of the seizure. One should try to classify the seizure as having a focal or generalised onset. In focal onset the seizure originates in one hemisphere of the brain. In the previous classification the term ‘partial’ was used but that has been replaced with the term ‘focal’. In generalised onset there is EEG and/or clinical indication that the onset of the seizure is within and rapidly engages a bilaterally distributed network. If the nature of the seizure onset is known with less than 80% confidence by the clinician, it should be classified as ‘unknown onset’. As new information becomes available it may be possible to change the classification to either focal or generalised.

The guidelines then allow for optional further classification.

Further classification for focal onset seizures

Is awareness retained?

Focal onset seizures are subdivided into those with retained awareness during the seizure (not necessarily awareness of the seizure itself) and those with impaired awareness during

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any part of the seizure. Focal aware seizures is the new term for 'simple partial seizures' in the 1981 classification and focal impaired awareness seizures replaces the previous term 'complex partial seizures'. The degree of consciousness throughout a seizure has a profound practical implication for forecasting the impact of the seizure on behaviour. It is important to note that recording awareness level need is not compulsory and may be omitted if it is unknown or unnecessary as in the case with focal myoclonic seizures.

Is the onset motor or non-motor?

The subsequent classification of focal onset seizures is motor vs. non-motor onset. The subtypes of these categories are described in detail in the expanded classification section.

Focal to bilateral tonic–clonic seizures

A seizure which begins focally and subsequently spreads to cause bilateral tonic–clonic movements is termed a 'focal to bilateral tonic–clonic seizure'. These used to be classified as 'secondary generalised tonic–clonic seizures', but in the 2017 classification the term 'generalized' is reserved to describe seizures that are generalised from onset. The new term is a clearer and more accurate descriptor. This is a common and important type of seizure.

Further classification of generalised onset seizures

Because the large majority of generalised onset seizures is associated with impaired awareness, awareness is not used as a classifier.

Generalised onset seizures are grouped into the two categories of motor and non-motor (absence - often termed 'petit mal' in the past) seizures. Motor activity should be bilateral from the onset, and can be recorded as tonic–clonic (often termed 'grand mal' seizures in the past) or other motor.

Further classification of unknown onset seizures

These can be classified as motor (tonic–clonic or other motor) or non-motor. If there is a dearth of information, 'unclassified' is allowed.

The expanded seizure classification

Focal onset seizures

Is awareness retained?

Although it is not mandatory to classify awareness for most seizures this remains very important. For the (relatively rare) atonic seizures and epileptic spasms, the expectation is that awareness will not be specified.

Focal onset seizures can be further subclassified by the addition of any motor onset or non-motor onset symptoms, corresponding to the initial principal characteristic displayed in the seizure, irrespective of whether it is the predominant symptom overall. This is important because determining the earliest feature can often indicate the seizure's neural origin.

Focal motor onset symptoms

Focal motor onset attributed behaviours encompass changes in certain aspects of movement activity. These can comprise automatisms, hyperkinesia, or other newer types which have formerly only been identified within generalised onset seizures, including atonic, tonic, clonic, myoclonic and epileptic spasms. It is important to note that although these types can appear in both focal and generalised onset seizures, they do not necessarily share the same pathophysiology or seizure prognosis. Automatisms are involuntary, repetitive and unconscious motor responses that can occur during seizure activity, such as repetitive tapping, mouth movements (e.g. lip-smacking) and repetitive hand movements (e.g. rubbing hands). Sometimes these can overlap with hyperkinetic seizures (or hypermotor seizures), which can involve pedalling, pelvic thrusting or thrashing movements. Therefore, this can cause some uncertainty in distinguishing between these types of seizures. Focal atonic seizures present with a sudden diminishing of limb tone, whereas focal tonic seizures involve the sudden stiffening and contraction of the limb or neck muscles. Clonic activity involves a constant regular rhythmic jerking or twitching whereas myoclonus is a transitory irregular jerking. Epileptic spasms (previously known as infantile spasms) are manifested as a sudden involuntary flexion, usually at waist level accompanied by splayed arm flexion or extension. Normally seen in young children between 3 and 8 months, they require monitoring of both the brain's electrical activity patterns via the EEG, and consideration of any merging clinical patterns, in order to fully differentiate between focal and generalised onset.

Focal non-motor onset symptoms

Focal non-motor onset symptoms involve alterations in emotion, sensation or cognition. These can encompass behavioural arrest seizures, described as a coordinated inhibition of movement and responsiveness, with the patient often observed as freezing. Due to transitory behaviour arrests being fairly common and challenging to pinpoint at the beginning of a seizure, a focal behaviour arrest seizure can only be classified as such if it is the prevalent characteristic of the seizure, and contrary to most cases, regardless of whether it was the earliest feature observed.

Other focal non-motor features include autonomic seizures, in which the main features are altered autonomic system functioning such as gastrointestinal sensations, palpitations and flushing. Focal cognitive seizures relate to when cognitive deficits are the primary symptoms that occur in a seizure. These can include language impairments, diminished spatial awareness or other higher cortical dysfunction. Such aberrant cognitive phenomena can be exhibited as forced thinking, hallucinations and déjà (or jamais) vu experiences. Focal emotional seizures usually manifest themselves during the seizure as an initial sudden outburst of emotion, such as fear, agitation, elation or paranoia, and may cause spontaneous laughter. Some of these emotions experienced by the patient may be subjective, and as such necessitate the individual to recount this, which can be problematic when awareness has been impaired. If this loss of awareness should occur, seizure

classification should not be assigned as a focal cognitive seizure, since diminishing awareness is applicable to all focal seizures. Focal sensory seizures entail disturbances in various sensory modalities including visual, auditory, gustatory, somatosensory and vestibular.

Generalised onset seizures

The 2017 classification of generalised onset seizures is similar to the 1981 classification, with the addition of some more types. They are grouped into generalised onset motor and non-motor (absence) seizures.

Motor onset

Generalised onset tonic–clonic seizures typically last less than 2 min. There is an immediate loss of awareness coupled with stiffening of all limbs (tonic phase), followed by sustained rhythmic jerking of limbs and face (clonic phase). A cry at the start, falling (backwards), tongue biting and incontinence may be associated features.

A generalised onset clonic seizure does not have a tonic phase but a bilateral and sustained rhythmical jerking of the limb and/or head. Such seizures usually occur in young children.

In generalised onset tonic seizures all the limbs stiffen; however, there is no clonic phase. Generalised onset myoclonic seizures are characterised by unsustained irregular bilateral jerking of limbs, face, eyes or eyelids.

The myoclonic-tonic–clonic type is new to the 2017 classification. This is an important addition as a few myoclonic jerks prior to the tonic and clonic phases are commonly seen in juvenile myoclonic epilepsy. A generalised myoclonic-atonic seizure manifests with a few myoclonic jerks which precede a limp drop. Atonic seizures tend to last for seconds and are an epileptic drop attack, with sudden loss of muscle tone and strength and a fall onto the buttocks, or forwards, This type typically occurs in children with severe epilepsy syndromes.

Generalised onset epileptic spasms are very short seizures in which there is flexion at the trunk accompanied by flexion or extension of the limbs. These were previously termed infantile spasms.

Non-motor onset

In a typical absence there is a sudden onset of activity cessation and occasionally automatic behaviours such as eye fluttering and head nodding. If this seizure type lasts for more than a few seconds, there is impairment in awareness and memory. There are characteristic EEG changes during typical absences (generalised spike and waves). Absences are common in children but rarely present in adulthood, and it should usually be possible to distinguish them from focal impaired awareness ('complex partial') seizures.

Atypical absence seizures are similar to typical absence seizures; however, they may have slower onset and recovery with more marked changes in tone. A difficulty with atypical absence seizures is that they can be difficult to differentiate from focal impaired awareness seizures. A generalised myoclonic absence seizure is defined as a seizure with a few jerks followed by an absence seizure. The jerks of the eyelids and upward deviation of the eyes, usually provoked by closing the eyes or by light, is called eyelid myoclonia. It is important to note that eyelid myoclonia may be associated with absence seizures.

Conclusion

The 2017 classification allows for more robust, specific and flexible classification of seizures. Some seizures (tonic, atonic, myoclonic, clonic, and epileptic spasms) that were previously only recognised as generalised onset now have focal counterparts. New focal seizure types that have been introduced include automatism, behaviour arrest, autonomic, cognitive, and emotional. New generalised seizure types include myoclonic-tonic–clonic, myoclonic absence, and absence with eyelid myoclonia. If the onset is unknown for tonic–clonic, behaviour arrest and epileptic spasms, they can be classified provisionally. The terminology has been simplified somewhat, avoiding confusing or unclear terms such as 'psychic', 'simple', 'complex', and 'dyscognitive,' and made more logical.

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