Epilepsy: epidemiology, classification and natural history

Heather Angus-Leppan
Linda M Parsons

Abstract

Epilepsy is a paroxysmal disorder in which a person has two or more seizures. A seizure results from an abnormal electrical discharge and the clinical manifestations vary greatly. The peak incidence is in younger and older people. Childhood febrile convulsions occur in about 5% of the population, and there is a 2–5% lifetime prevalence for one or more seizures. Two-thirds of patients go into remission. The minority are refractory and need reassessment of both the diagnosis and management. Epilepsy is associated with at least twice the standardized mortality ratio. In the economically disadvantaged world epilepsy is even more common, compounded by far fewer resources to deal with it. The different patterns and causation are summarized.

Keywords classification; cryptogenic; epilepsy; focal; idiopathic; primary; SUDEP; symptomatic

Definition

Epilepsy means two or more seizures. Hughlings Jackson defined seizure manifestations as due to an occasional, sudden, excessive, rapid and disorderly discharge in part of the cerebral cortex.[1] This definition still applies.

Seizure manifestations range from brief sensory experience, to microsecond lapses in concentration, to convulsive status epilepticus. Aetiology, associated morbidity, natural history and prognosis differ. Some writers prefer 'the epilepsies' to 'epilepsy' to highlight this variability but the term 'epilepsy' does convey important shared features:

- paroxysmal disturbance
- return to status quo between episodes
- the underlying mechanism of disturbed cerebral electrical rhythms.

Terminology

Important terms are summarized in Table 1. Those with a specific meaning in epileptology are marked with an asterisk.

Historical perspective

Early records of epilepsy from 1000BC[1] suggested it was due to demons, misdeeds, magical or astrological influence. This concept still has an impact on lay views of epilepsy. From Hippocrates (400 BC) others viewed epilepsy as having a natural but unknown cause.[1] Hughlings Jackson viewed epilepsy as the result of abnormal cerebral discharges. This paradigm shift in the 1800s built on Ferrier and Todd's experiments[2] at the time when the first modern treatment for epilepsy (potassium bromide) further stimulated interest in the subject. Since then epileptology has expanded on Jackson's concepts and our range of therapies.[1]

Epidemiology

The epidemiology of epilepsy is imprecise due to variations in methodology, inclusion criteria and diagnostic uncertainty. Epilepsy starts at any age, with peak incidences in early and late life (Figure 1).

Incidence

The incidence of epilepsy in the economically developed world, excluding febrile convulsions and isolated seizures, is estimated at 50/100,000/year (range of 40–70/100,000).[3] In the developing world, epilepsy is probably twice as common due to infection, social problems and economic deprivation.

Prevalence

This is estimated as 5–10/1000, excluding epilepsy in remission (inactive epilepsy), febrile convulsions and single seizures.[3] The lifetime prevalence of a seizure (excluding febrile seizures) is 2–5%.

Approximately 67 million people in the world have epilepsy, 600,000 in the UK. In a primary-care British practice with 2500 patients, about 13–25 will have active epilepsy, and one to three will have severe epilepsy with associated major handicap.[4]

Classification

Seizures

The classification of seizures is summarized in Table 2, and terms used in Table 1. Many manifestations are seen in different types of epilepsy and are not specific. For example, simple partial and tonic–clonic seizures may occur in one person due to focal epilepsy, such as due to a tumour. Some epilepsy syndromes have more than one seizure manifestation. For example, juvenile myoclonic epilepsy may manifest as myoclonus, absences and convulsions. The main seizure types are summarized below.

Heather Angus-Leppan *MSc (Ep) MD FRACP FRCP is a Consultant Neurologist, Honorary Senior Lecturer and Epilepsy Lead at the Royal Free Hospital, Pond Street, London, UK. She is the Honorary Assistant Secretary of the Association of British Neurologists, Honorary Secretary of the Neurosciences Section of the Royal Society of Medicine and current Chair of the Map of Medicine Epilepsy Group, UK. Competing interests: none declared.*

Linda M Parsons *MD FRCP is a Consultant Neurologist at Royal Free, Luton and Dunstable and St Albans City Hospitals, UK. She qualified from Sheffield University and trained in Neurology in Birmingham and London, UK. She has a longstanding interest in epilepsy and currently sits on the steering committee for the UK Epilepsy and Pregnancy Register. Competing interests: none declared.*

Terminology

Aura	Subjective experience as a result of seizure discharge. It can occur alone (a simple partial seizure) or be followed by other seizure manifestations. An aura is a seizure, not a prodrome to a seizure.
Automatism	Complex automatic behaviour associated with loss of awareness, such as lip smacking or hand wringing, occurring as part of a seizure.
Convulsion*	Term used for a seizure of any type with involuntary, irregular myoclonic, clonic or tonic–clonic movements of one or more limbs. It is most applicable to the form of generalized seizure with tonic–clonic movements (*grand mal*), emphasizing the generalized 'convulsive' type, as opposed to generalized 'non-convulsive' (e.g. childhood absence) epilepsy.
Cortical malformations	These are also referred to as *cortical dysgenesis*, *cortical dysplasia* or *neuronal migration disorders* and are developmental abnormalities of the cerebral cortex.
Cryptogenic*	Cause unknown, presumed symptomatic.
Déjà vu	Literally 'already seen', this refers to a false impression that a present experience is familiar. It is used to refer to something heard, experienced or seen. It can be the aura of a temporal lobe seizure, although it also happens in other settings.
Epilepsy	Two or more seizures.
Epilepsy in remission	No seizures for 2–5 years.
Fit	A popular lay term to denote a seizure of any type but also an outburst of temper or aggression.
Ictus	Literally a 'blow'. Usually used for an epileptic seizure (but can refer to other paroxysmal events, such as migraine, transient neurological events and stroke).
Idiopathic (primary)*	In relation to epilepsy, this implies that there is an underlying genetic cause.
Incidente	The rate of an event in a population over a certain period of time.
Jamais vu	Literally 'never seen', this refers to a false impression that a present experience is unfamiliar. It is used for something seen, heard or experienced. It can be the aura of a temporal lobe seizure, although it also happens in other settings.
Learning disability	Difficulties with daily living associated with intelligence quota less than 70 and age of onset less than 18 years.
Major seizure	A loose term that refers to a generalized convulsive seizure.
Minor seizure	A loose term that refers to seizures that are not generalized convulsive.
Partial (focal, localization related) seizure	A seizure originating in a specific cortical location.
People with epilepsy (PWE)	Term used instead of 'epileptics' to emphasize the whole person and that epilepsy is a condition someone has, and not their identity.
Postictal	The period of time after a seizure when behaviour or mood can be directly affected by the seizure.
Prevalence	The rate of an event in a population at a single point in time.
Primary generalized seizure	A seizure involving both hemispheres from onset.
Refractory epilepsy	Not responding to appropriate medical treatment over at least 2 years.
Seizure	Clinical manifestation of a sudden abnormal cerebral electrical discharge.
Standardized mortality ratio	The ratio of the observed deaths in a study population to the expected number experienced by the age- and sex-matched control group.
Sudden unexplained death in epilepsy (SUDEP)	Sudden, unexpected death in a person with epilepsy not due to drowning or other accident; without or with a seizure. Deaths during status epilepticus or those with an anatomical or toxicological explanation are excluded.
Symptomatic epilepsy	Having an identified underlying cause.
Syndrome	A constellation of clinical and investigational features for which there may be a number of aetiologies.
Typical absence (petit mal)	A generalized seizure characterized by loss of awareness, usually brief and associated with characteristic 3–4/second spike and wave discharges on an electroencephalogram. This must be distinguished from atypical absence and complex partial seizure.

*These terms have acquired a specific meaning in epilepsy different to their use in other fields of medicine.

Table 1