# Paper 1: Defining epilepsy

## Epilepsy

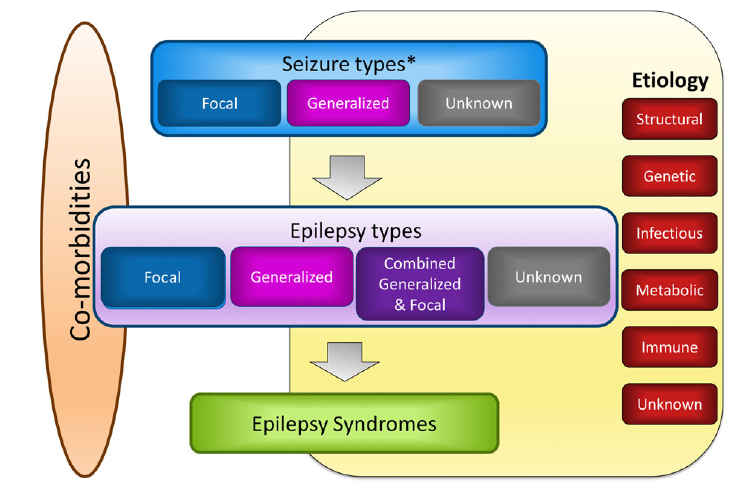
Epilepsy is a condition defined by the occurrence of epileptic seizures. Epileptic seizures are events that arise due to abnormal electrical activity in the brain. These epileptic seizures can range from brief behavioral arrests to stiffening and or jerking of the whole body. The International League Against Epilepsy (ILAE) defines epilepsy as:

* at least two unprovoked (or reflex) epileptic seizures occurring more than 24 hours apart
* one unprovoked (or reflex) epileptic seizure, and a probability of further epileptic seizures similar to the general recurrence risk (at least 60 percent) after two unprovoked seizures, occurring over the next 10 years
* diagnosis of an epilepsy syndrome.

Epilepsy is considered to be resolved for individuals who had an age-dependent epilepsy syndrome but are now past the applicable age, or those who have remained seizure-free for the last 10 years with no seizure medicines for the last five years (International League Against Epilepsy Commission, 2014).

Epilepsy is a group of disorders with varying seizure types, age of onset, severity and long term outcome. It can present at any age but has peaks of onset in infancy/childhood and the elderly. There are multiple causes of epilepsy. Most individuals with epilepsy in New Zealand have an underlying genetic cause. This can be due to an abnormality in single gene or due to the interaction of multiple genes and is often not inherited. Epilepsy can also be acquired due to an insult to the brain such as head injury, hypoxic ischaemic injury (including stroke), infection or immunologically mediated disorders (Sheffer, et al. 2017, Fisher, et al. 2017).

Figure 1: Classification of the epilepsies



Source: Scheffer et al. 2017, p4

The number of people affected by epilepsy in New Zealand is not clearly defined. International epidemiological studies have estimated the incidence of epilepsy to be from 40−77 per 100,000 (of population) however this does vary depending on the study cited (Neligan 2011). The incidence of childhood epilepsy (number of new cases) has been reported to be higher than the adult population.

## Drug-resistant epilepsy

Although the majority of people with epilepsy will become seizure free withanti-epileptic drugs (AEDs), up to 30 percent have drug-resistant epilepsy. Drug-resistant epilepsy is defined as ‘a failure of adequate trials of two tolerated and appropriately chosen and used anti-epilepsy drug schedules (whether as mono therapies or in combination) to achieve sustained seizure freedom’ (Kwan 2010).

Patients with drug-resistant epilepsy are more likely to have cognitive, psychological and social co-morbidities than those who are seizure free.

There are a significant group of individuals with drug-resistant epilepsy who will benefit from epilepsy surgery. Epilepsy surgery includes:

1. resection of an epileptic focus
2. disconnection of an epileptic focus
3. implantation of a device that provides electrical stimulation to the brain or a peripheral nerve.

## Status epilepticus

The ILAE have recently redefined tonic-clonic status epilepticus as:

“a condition resulting either from the failure of the mechanisms responsible for seizure termination or from the initiation of mechanisms, which lead to abnormally, prolonged seizures (after 5 min). It is a condition, which can have long-term consequences (after time 30 min), including neuronal death, neuronal injury, and alteration of neuronal networks (Trinka, et al. 2015).”

Previously the definition of status was a single seizure of more than 30 minutes duration or a series of epileptic seizures during which function is not regained between ictal events in a 30 minute period (International League Against Epilepsy 1993).

## Sudden unexpected death from epilepsy (SUDEP)

People with epilepsy have a small risk of SUDEP. SUDEP is defined as:

“the sudden, unexpected, witnessed or unwitnessed, non-traumatic and non-drowning death in patients with epilepsy, with or without evidence for a seizure, and excluding documented status epilepticus, in which post-mortem examination does not reveal a structural or toxicological cause of death (Nashef, So, Ryvlin, & Tomson 2012)”

SUDEP is reported to be the cause of 2-18 percent of all deaths in people with epilepsy, and potentially as high as 30 percent of deaths in children with epilepsy (Schachter 2016). The overall incidence is 1 in 4500 patient years in children and 1 in 1000 patient years in adults (Harden, et al. 2017).

The cause of SUDEP is unknown, but is theorised to be due to respiratory suppression followed by cardiac arrest in the postictal period of a tonic-clonic seizure. The main risk factor is the presence of tonic-clonic seizures. One or two tonic-clonic seizures per year increases the risk 5 fold while the presence of more than 3 tonic-clonic seizures per year results in a 15 fold increased risk (Harden, et al. 2017). Strategies for preventing SUDEP are therefore linked to prevention of tonic-clonic seizures.

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