Paper 1: Defining epilepsy

Epilepsy

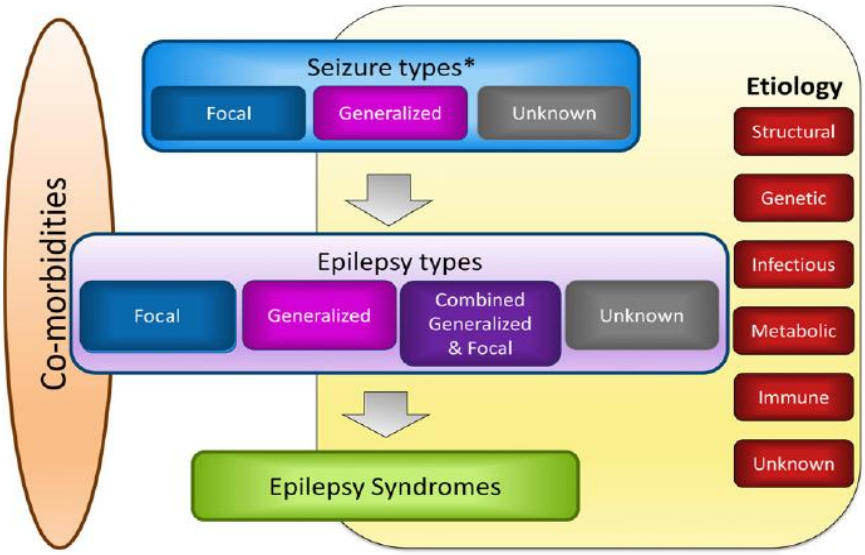
Epilepsy is a condition defined by the occurrence of epileptic seizures. Epileptic seizures are events that arise due to abnormal electrical activity in the brain. These epileptic seizures can range from brief behavioral arrests to stiffening and or jerking of the whole body. The International League 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 with anti-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.

References

International League Against Epilepsy. (1993). Guidelines for epidemiological studies on epilepsy. *Epilepsia*, 592.

Baille, C. (2011). *Epilepsy and the workplace: a guide for workers and employers*. . Canberra: The Epilepsy Association of the ACT Inc.

Bergin P, S. L. (2008). Bringing epilepsy out of the shadows in New Zealand. *New Zealand Medical Journal, vol 121, no 1268*, URL: http://www.nzma.org.nz/journal/121-1268/2893/.

Boon, A. (2012). *Excellence through Patient and Family Centred Care*. New Zealand: Health Quality Safety Commission.

Booth, L., & Thompson, G. (2010). *Epilepsy Statistics*. London: Social and General Statistics.

Bradley, P., & Lindsay, B. (2008). Care delivery and self management strategies for adults with epilepsy. *Cochrane database of systemic reviews*, Issue 1, art no. CD006244; DOI: 10.1002/14651858.

Burkett, I. (2016). Co-design for social good. *Rethinking Aging*. Sydey: Design 4 social innovation.

Craig, E. R. (2013). *The Health of Children and Young People with Chronic Conditions and Disabilities in the South Island*. Christchurch: South Island Alliance. Retrieved from www.sialliance.health.nz/.../SouthIslandAlliance/.../3_%20Introduction_.

Department of Health. (2008). *Western Australia: Epilepsy Model of Care*. Perth: Health Networks Branch, Department of Health, Western Australia.

Drislane, F. W. (2017, April 04). *Convulsive status epilepticus in adults: treatment and prognosis*. Retrieved from UpToDate: https://www.uptodate.com/contents/convulsive-status-epilepticus-in-adults-treatment-and-prognosis?source=search_result&search=status%20epilepticus&selectedTitle=1~150#H2329061529

Epilepsy Association of New Zealand. (2016, October). *The Workplace and Epilepsy: a guide for New Zealand workers and employers*. Retrieved from Epilepsy New Zealand: http://epilepsy.org.nz/files/New%20Zealand%20Employment%20and%20Epilepsy%20Report%202016%20Final(2).pdf

Epilepsy Foundation. (2013, October). Retrieved August 25, 2016, from http://www.epilepsy.com/learn/epilepsy-statistics

Epilepsy Organisation of Australia. (2016). *Epliepsy Organisation / Resources / Facts and statistics about epilepsy*. Retrieved August 2016 25 August 2016, 2016, from Epilepsy

Orgnaisation: https://www.epilepsy.org.au/resources/for-media/facts-statistics-about-epilepsy

Fitzsimons, M., Normand, C., Varley, J., & Delanty, N. (2012). Evidence-based models of care for people with epilepsy. *Epilepsy & Behaviour*, Vol 23, Issue 1, pp1-6.

Hackett, M. L., Glozier, N. S., Martiniuk, A. L., Jan, S., & Anderson, C. S. (2011). Sydney epilepsy incidence study to measure illness consequences: the SEISIMIC observational epilepsy study protocol. *BMC Neurology*.

Health Service Executive. (2012). *The National Epilepsy Care Programme in Ireland*. Dublin: Health Services Executive, Clinical Strategy and Programmes Directorate.

Health Service Executive. (2016). *The National Clinical Programme for Epilepsy - Model of Care*. Dublin: Health Services Executive, Clinical Strategy and Programmes Directorate.

International League Against Epilepsy. (1993). Guidelines for epidemiological studies on epilepsy. *Epilepsia*, 592.

International League Against Epilepsy Commission. (2014). *http://www.ilae.org/visitors/centre/Definition-2014.cfm*.

Joint Epilepsy Council of the UK and Ireland. (2011). *Epilepsy prevalence, incidence and other statistics*. London: Joint Epilepsy Council.

Kwan, P., Arzumanoglou, A., Berg, A. T., Brodie, M. J., Hauser, W. A., Mathern, G., . . . French, J. (2010). Definition of drug resistant epilepsy: Consensus proposal by the ad hoc Task Force of the ILAE Commission on Therapeutic Strategies. *Epilepsia*, 1069-1077.

Ministry of Health. (2012). *National Services: Service Review Complex Epilepsy*. Wellington: Ministry of Health.

Ministry of Health. (2015). *Disability Support Services Strategic Plan 2014 to 2018*. Wellington: Ministry of Health.

Ministry of Health. (2016). *Self-management support for people with long term condtions*. Wellington: Ministry of Health.

Ministry of Health. (2016). *The New Zealand Health Strategy*.

Ministry of Health. (2017). *Health and Independence Report 2016. The Director-General of Health's Annual Report on the State of Public Health*. Wellington: Ministry of Health.

Nashef, L., So, E. L., Ryvlin, P., & Tomson, T. (2012, 53(2)). Unifying the definitions of sudden unexpected death in epilepsy. *Epilepsia*, 227.

National Health Committee. (2007). *Meeting the needs of people with chronic conditions*. Wellington: National Advisory Committee on Health and Disability.

Neligan, A. (2011). *The Long Term Prognosis of Epilepsy*. London: University College of London.

Neligan, A., & Sander, J. W. (2009). The incidence and prevalence of epilepsy. In J. a.-G. Sander, *Epilepsy 2009: from benchside to bedside. A practical guide to epilepsy*.

Lecture notes from the Twelfth Epilepsy Teaching Weekend (pp. 15-21). Oxford: UCL Institute of Neurology.

Ngugi, A. K., Kariuki, S. M., Bottomley, C., Kleinschmidt, L., Sander, J. W., & Newton, C. R. (2011). Incidence of epilepsy: a systematic review and meta-analysis. *Neurology, 77(10)*, 1005-1012.

NICE. (2016, November). *Epilepsy*. Retrieved from National Institute for Health and Care Excellence: https://www.nice.org.uk/guidance/conditions-and-diseases/neurological-conditions/epilepsy

Nixon, A., Kerr, C., Breheny, K., & Wild, D. (2013). Patient Reported Outcome (PRO) assessment in epilepsy: a review of epilepsy-specific PROs according to the Food and Drug Aministration regulatory requirements. *Health and Quality of Life Outcomes*, 11:38.

Salpekar J, B. M. (2013). Epidemiology and common comorbidities of epilepsy in childhood. In C. D. Wheless J, *Epilepsy in Children and Adolescents*. West Sussex: Wiley-Blackwell.

Schachter, S. C. (2016, December 06). *Management of epilepsy and pregnancy*. Retrieved from UpToDate: https://www.uptodate.com/contents/management-of-epilepsy-and-pregnancy?source=search_result&search=epilepsy%20pregnancy&selectedTitle=1~150

Schachter, S. C. (2016, November 01). *Sudden unexpected death in epilepsy*. Retrieved from UpToDate: https://www.uptodate.com/contents/sudden-unexpected-death-in-epilepsy?source=search_result&search=sudep&selectedTitle=1~14

Shackleton, D., Kasteleijn-Nolst Trenite, G., de Craen, J., Vandenbroucke, J., & Westendorp, R. (2003). Living with epilepsy. Long-term prognosis and psychosocial outcomes. *Neurology, Vol 61, No. 1*, 64-70.

SIGN. (2015, May). *Diagnosis and management of epilepsy in adults*. Retrieved from Scottish Intercollegiate Guidelines Network (SIGN): www.sign.ac.uk

Statistics New Zealand. (2015). Wellington: Statistics New Zealand.

Technical Advisory Group. (2012). *Service Review: Complex Epilepsy Services in New Zealand*. Wellington: Ministry of Health.

Wilfong, A. (2016, September 30). Seizures and eiplepsy in children: classification, etiology, and clinical features.

Wong, A. (2015, November 16). *Clinical features and complications of status epilepticus in children*. Retrieved from UpToDate: https://www.uptodate.com/contents/clinical-features-and-complications-of-status-epilepticus-in-children?source=search_result&search=status%20epilepticus&selectedTitle=4~150#H20

World Health Organisation. (2005). *Atlas: Epilepsy care in the world*. Geneva: World health Organisation.

World Health Organisation. (2016). *Fact Sheet - Epilepsy*. Geneva: World Health Organisation.

Kwan, A. A. (2010). Definition of drug resistant epilepsy: Consensus proposal by the ad hoc Task Force of the ILAE Commission on Therapeutic Strategies. *Epilepsia*, 1069-1077.

Nashef, L., So, E. L., Ryvlin, P., & Tomson, T. (2012, 53(2)). Unifying the definitions of sudden unexpected death in epilepsy. *Epilepsia*, 227.

Neligan, A. (2011). *The Long Term Prognosis of Epilepsy*. London: University College of London.

Salpekar J, B. M. (2013). Epidemiology and common comorbidities of epilepsy in childhood. In C. D. Wheless J, *Epilepsy in Children and Adolescents*. West Sussex: Wiley-Blackwell. .

Schachter, S. C. (2016, November 01). *Sudden unexpected death in epilepsy*. Retrieved from UpToDate: https://www.uptodate.com/contents/sudden-unexpected-death-in-epilepsy?source=search_result&search=sudep&selectedTitle=1~14