1

What is epilepsy?

Key points

- The human brain has over 100,000 million nerve cells.
- Epileptic seizures occur when nerve cells fire abnormally and synchronously.
- Not all seizures are due to epilepsy.
- There are numerous types of seizures.
- There are many potential causes of epilepsy.
- Epilepsy is diagnosed after two or more seizures.
- Epilepsy is common, affecting 3–4 per cent of people by the time they reach the age of 75.
- Epileptic seizures will stop with drug treatment for at least 60 per cent of people.

How nerve cells work

The human brain contains about 100,000 million nerve cells, each of which is connected to many others, perhaps as many as 50,000 others. The brain is the organ of our thinking, memory and emotion. It integrates information from the outside world and so allows us to perceive objects, such as those we can see, hear, touch or smell, and enables us to perceive events around us. It organizes our response to these events by movements or other actions. It organizes our social behaviour.

Messages are passed between nerve cells by the extraordinarily rapid secretion of tiny packets of specialized chemicals known as neurotransmitters. As a neurotransmitter acts on the next cell in a chain, a brief electric current is generated. These can be recorded by very fine wires placed next to or in a nerve cell, but they are not large enough to be recorded externally over the skin of the head. However, some cells act in rhythmic concert, and these rhythms can be detected with devices such as the electroencephalogram (EEG), a test in which electrodes are placed over the scalp and these electrical rhythms are amplified and recorded digitally or on tape or disc, and displayed on a computer screen, or rarely, a moving strip of paper. The use of this procedure is described in Chapter 5.

Some messages received by a nerve cell are inhibitory—they dampen down the activity of the receiving cell. Some are excitatory, enhancing its activity. The receiving nerve cell computes, as it were, these contrasting messages, which determine its own action.

The events leading to a seizure

One of the ways in which events can go wrong is when a nerve cell loses some of its inputs from other cells because of damage to these other nerve cells. If inhibitory terminals are lost, then the cell will become over-excitable, and begin to switch on, or fire inappropriately, driving other nerve cells with which it is connected. This may result in more and more nerve cells being incorporated into the abnormal pattern of firing.

Thus during an epileptic seizure the normal, quiet, and integrated function of nerve cells in the cerebral hemispheres is interrupted. Instead, nerve cells are forced, through the contacts they make with and receive from other nerve cells, into an abnormal firing pattern, and as the seizure develops, more and more nerve cells become involved. These nerve fibres fire synchronously in paroxysms.

There are a number of different types of seizures, and these are a reflection of different patterns of paroxysmal discharge of nerve cells. Seizure types are described in Chapter 2 and two examples are given here. If the seizure discharge spreads throughout large areas of the brain, then consciousness may be lost. If the discharge of nerve cells is confined to one small area, such as to part of the temporal lobe of the brain (more or less above and in front of the ears), the person may remain conscious but experience a distortion of memory so that the person perceives that he or she has experienced ongoing events before - the phenomenon of déjà vu.

The definition of an epileptic seizure

It is difficult to give a short definition of a seizure, due to the different types of seizure that may occur and the range of symptoms that may be experienced by the person having a seizure, and the range of features that may be observed. An epileptic seizure occurs when there is an abnormal paroxysmal discharge of nerve cells in the cerebral cortex that causes symptoms for the person and/or changes apparent to an observer. The precise symptoms and changes will depend on where in the brain the seizure starts where and how far it spreads. For example, a seizure starting in the temporal lobe may start with a sensation of déjà vu. A seizure rapidly spreading to both cerebral hemispheres might have no warning symptoms, but an observer might see the person go stiff and then shake all four limbs (a convulsion).

In someone with established epilepsy, the EEG between seizures may also show abnormal discharges which are not apparent to the doctor in terms of observed behaviour, nor are they associated with any change perceived by the person with epilepsy. Although the abnormal discharges of the EEG are clearly a fragment, as it were, of a seizure, they are not usually regarded as seizures, as no change is noticed by the person or observer.

The definition of epilepsy

Not all seizures are due to epilepsy. For example, some poisons (or drugs) might cause a person to have a seizure, and similarly an alcoholic might have a seizure whilst withdrawing from alcohol. Without the poison or alcohol withdrawal, these individuals would not have seizures, and they would not be said to have epilepsy. A person is said to have epilepsy if they have recurring seizures (more than two) for which no such provoking factor is found.

There are, however, many grey areas which can cause confusion, some of which are discussed later in this book. Take for example the case of a young man who has a single seizure at the age of 19, the day following a late night and heavy alcohol intake. It would be justifiable to assume that alcohol played some part in the cause of the seizure, but there were others who drank just as much who did not have a seizure. So we must presume that the man has a lower seizure threshold than his friends (see Chapter 3).

A single seizure is not considered sufficient to make the diagnosis of epilepsy, and a diagnosis of epilepsy is usually made following two or more seizures. Around one half of people who have a first seizure will go on to have a second one over the next three years.

Again, there may be grey areas in applying this definition of epilepsy to everyone who has had two seizures. Consider a man who had a seizure at the age of 19 and another at the age of 80. It would seem a bit nonsensical to tell the elderly man that he had had epilepsy all his life, as we would be obliged to do if we followed this definition rigidly. Similarly, consider a woman aged 40, who had ten seizures between the ages of 15 and 25. Although her epilepsy might be considered to still be in remission, she remains at a small risk of having a seizure. Should we consider her as someone with epilepsy? These examples hopefully show that the label 'epilepsy' has to be applied with common sense. It is not one of those tidy diseases such as myocardial infarction, in which there is little argument about the heart attack or the coronary disease causing it.

These medical uncertainties and complexities can make it difficult to give a crystal clear definition and explanation of the illness. Epilepsy is a common condition that has been recognized since ancient times, and there is a common folklore around epilepsy which is full of half truths and inaccuracies. Half truths, include the notion that epilepsy always starts in childhood, is inherited, and is always convulsive in nature. Inaccuracies include the notions that epilepsy is a mental illness and that women with epilepsy cannot have children. Some of these half truths and misconceptions can be a cause of major stigma to people with epilepsy. In this book, we hope to provide some clarity about epilepsy and dispel these ideas.

Identifying the cause of epilepsy

Once a doctor has decided that a person has epilepsy, the next question to address is usually, why? There are many, many potential causes of epilepsy, and the doctor will usually have a good idea as to the cause according to the types of seizure and the age of the person. For up to two-thirds of the people, no specific cause is found, and for many of these the cause is thought be genetic; epilepsies with a genetic cause most commonly start in childhood and adolescence. If the doctor thinks that the seizures are unlikely to be genetic in origin, a brain scan—computerized tomography (CT) or magnetic resonance imaging (MRI)—is usually undertaken to try and identify a brain abnormality that is causing the epilepsy. On average, serious causes such as brain tumours are only found in around one in a hundred people.

Words used to describe epileptic seizures

Doctors and patients use many different words to describe seizures. The most readily recognized seizure in which a patient collapses, goes rigid and then had rhythmic shaking of all four limbs might be called a convulsion, a fit,

a grand mal attack, a generalized seizure, or a tonic–clonic seizure. The latter, tonic–clonic seizure, is the preferred medical term and will be used throughout this book. The different types of seizures are described in more detail later in this book. Throughout this book we use the word 'seizure' to refer to all seizures types except febrile convulsions, which are described in Chapter 9. In clinical practice, doctors will often use the words used by their patients such as fit, turn, attack or dizzy spell. People who have two types of seizure often call them 'big ones' and 'little ones'. As long as the patient and the doctor find themselves talking about the same events, this is perfectly acceptable.

Sometimes in correspondence and conversation doctors employ the words epileptiform or epileptoid. In our experience, doctors who use such terms are skating round the subject and avoiding frankly stating that their patient has had an epileptic seizure.

How common is epilepsy?

The incidence of a disease is a measure of the number of new cases in a defined population (usually 100,000) in a defined period of time (usually one year).

There have been a number of good research studies that have estimated the incidence of new cases of epilepsy, one of which was a study of the population of Olmstead County in Minnesota. People in this rural part of the USA do not move around very much, and have the good fortune to be cared for by doctors at the famous Mayo Clinic.

Research workers there have long had an interest in identifying all patients with epilepsy. Figure 1.1 shows the incidence of new cases of epilepsy (more than one non-febrile seizure) per 100,000 per year plotted against the age of onset. The incidence of new cases is highest in infancy and in old age for reasons which are explained in Chapter 3, but new cases can occur at any age. Throughout middle life the incidence is about 40 cases per 100,000 per year. As the years go by, the risk of having had epilepsy at some time in one's life increases in a cumulative fashion. From the United States study cited above, the cumulative risk by age 75 was 3,400 per 100,000 (3.4 per cent) for males and 2,800 per 100,000 (2.8 per cent) for females. In other words, anyone reaching the age of 75 has a 3–4 per cent chance of having had a diagnosis of epilepsy. Epilepsy is thus not a rare or unusual disorder; seizures may impinge upon the lives of anyone of us.

Prevalence is another measure used to assess how common a disease is. This is a measure of how many people in the population actually have a disease at

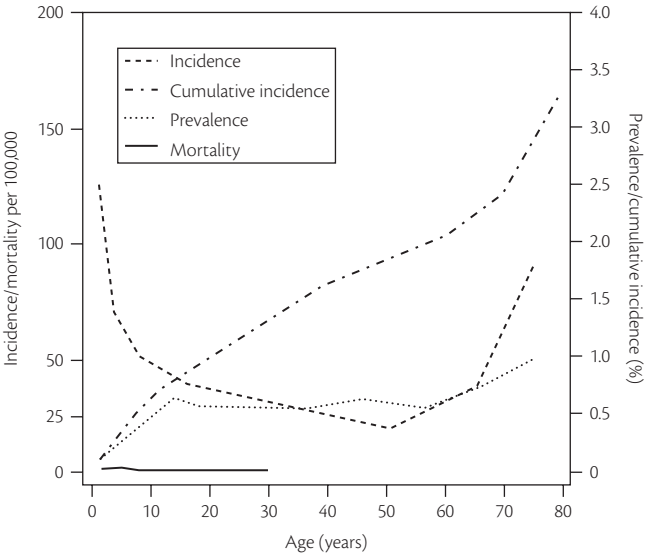

Figure 1.1 Incidence and cumulative incidence, prevalence and mortality rates for epilepsy in the Rochester, Minnesota (1935–1974) study. With permission from Anderson VE, Howser WA, Rich S.

any particular point in time. Prevalance is usually expressed as the number of cases per 1,000 population. Studying prevalence requires methods that are time-consuming and expensive, such as door to door surveys or in-depth study of all potential cases registered with general practitioners. Studying the prevalence of epilepsy is also complicated by a need to decide who should be classified as having ongoing or 'active' epilepsy. The sort of definition used typically is anyone who has had two or more seizures in their life, has had a seizure in the past two years and/or is taking anti-epileptic drug treatment. Estimates of prevalence around the world vary, but typical estimates for the developed world are 0.5 to 1 per cent. In other words, at any one time between 0.5 per cent and 1 per cent of the population have epilepsy. This again indicates that epilepsy is a common condition.

As seen in Figure 1.1 epilepsy may often start in the elderly. In western societies, people are living longer and there are more elderly people than ever before, which obviously means that there will be more elderly people with epilepsy. Recent research has even suggested that as a result of this, in some countries, it is now more common for epilepsy to start in the elderly than in children. However, it must also be emphasized that it is often difficult to

diagnose epileptic seizures in the elderly. The elderly are more likely to have multiple medical problems and might have falls or blackouts for a number of reasons and a careful history should be taken to help differentiate epileptic seizures from other events. For example blackouts may occur due to a heart problem, or due to a side-effect of treatment for another condition including hypertension or hypotension. Getting a good and accurate description from an eye-witness is essential, but sometimes made difficulty by the fact that increasing numbers of elderly people live alone following the death of their partner.

Does epilepsy stop?

There is one encouraging point that all those with epilepsy must remember: with anti-epileptic drug treatment, seizures stop for around 60 per cent of people. For the majority, seizures will be controlled in the first 1–2 years after starting treatment although for some there may be a period of attempting to find the right treatment or right combination of treatments before seizures may be controlled. Sometimes, surgery may help put seizures into remission. The chances of success with drugs or surgery depend in part upon the cause of the epilepsy and this is discussed in more detail in Chapter 6. For patients whose seizures have stopped, around half can stop treatment without seizures recurring. However, it is difficult to predict precisely who will have a recurrence and who will not, and many people decide to continue taking treatment rather than risk coming off treatment.