Diagnosis and Management of Epilepsy in the Adult Patient

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In the past 20 years there have been considerable advances in the understanding of the many problems related to epileptic seizures and of the difficulties which beset those who suffer from them. These advances have undoubtedly led to a general raising of the standards of treatment and management. There has also been some improvement in public attitudes towards people with epilepsy and quite recently attention has been drawn increasingly towards the ways and means by which its development might be prevented.

Nevertheless, there is still a great deal about epilepsy which is either unknown or only incompletely understood. However, it is not so much my task to concern myself with the unknown but rather to discuss briefly some of the more generally accepted principles applicable to the diagnosis and management of epilepsy in the adult patient.

DEFINITION OF EPILEPSY

I have long felt that it is misleading to speak of epilepsy as if it were a disease entity in its own right. On the contrary, I believe that it should be regarded as a symptom, just as we might regard a complaint of headache or shortness of breath, and that it must always have a cause even though it may not be immediately discernible.

Epilepsy is, in fact, the name we give to a particular kind of brief interruption of function in the brain. This is variously referred to as a fit or seizure, and when there is a continuing tendency for such episodes to recur we say a person so affected is suffering from epilepsy.

It has long been generally accepted that the basis of an epileptic seizure is a sudden excessive discharge of energy (or impulses) by groups of nerve cells in the brain. Such discharges are accompanied by chemical and electrical changes and cause temporary cessation or distortion of the normal functioning of the area of the brain so involved. If the area is localised to a relatively small segment of grey matter in one or other cerebral hemisphere, there results a cortical, focal or partial seizure; on the other hand, if it involves the deep, centrally placed nuclear masses of the brain (subcortical region or mesodiencephalon) the result is a generalised attack in which the seizure discharge involves the entire brain and in which consciousness is usually interrupted.

From these considerations it will be evident that the form which any given seizure will take is determined primarily by the site of origin of the discharge in the brain and by the particular function which the area affected subserves. Seizure discharge, however, has a tendency to spread beyond its area of origin and often, therefore, the pattern of the attack is also determined by the direction and rate of such spread. Bearing in mind the complexity and diversity of function in the brain, it can be no surprise that the different patterns of epileptic seizure occurring in human beings are very numerous. However, broadly speaking, these are likely to be characterised by motor or sensory manifestations (or both) and these are quite often, but by no means necessarily, associated with alterations in consciousness and awareness. Unfortunately, however, seizure discharge is very liable to involve those deeply situated areas of brain concerned with the maintenance of consciousness.

CLASSIFICATION OF SEIZURES

There are many different ways in which epileptic seizures may be classified. For example, they may be grouped according to the age of onset, the underlying cause, the form which they take, the nature of any precipitating factor or stimulus, whether or not consciousness is affected, and so on. Probably, however, one of the most widely accepted methods today is to divide seizures into two main categories—firstly, those which are generalised from the start, and secondly, those which are primarily of localised (cortical) origin, even though they may become generalised as a result of spread of discharge to the mesodiencephalic region of the brain (secondary generalisation).

Examples of primarily generalised seizures are petit mal and grand mal attacks; myoclonic episodes and most epileptic automatisms fall into the same category. Cortical seizures, on the other hand, show a much wider variety of patterns which may be broadly classified as motor, sensory and psychical. The latter include complex hallucinatory experiences which are, in fact, almost always the outcome of seizure discharges in certain parts of one or other temporal lobe of the brain.

UNIVERSAL OCCURRENCE OF EPILEPSY

The tendency for nerve cells in the brain to discharge energy in the manner described is a potential property

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which is inherent in everyone. Normally this is prevented from happening by certain inbuilt mechanisms having an inhibitory effect. In the very young these are not apparently fully operative, but in the adult they perform their role very effectively throughout life, unless they are damaged or the brain is subjected to highly abnormal stresses such as an electric shock of appropriate intensity. Even so, it has been estimated that something like 1 person in every 20 will have a seizure during the course of a lifetime, although only 1 in 8 of these will go on to have further attacks in the future.

There are many different circumstances under which epileptic seizures may occur in human beings. For practical purposes they may be classified under 3 main headings:

1. When brain cells have been damaged by injury or disease—as for example by such agents as physical injury, oxygen deprivation, infections, parasitic infestations, degenerative diseases, growths.

2. As a result of exposure to certain specific stresses in predisposed individuals—such as fevers (febrile convulsions), a flickering light (television epilepsy), sounds (including music), reading, movement.

3. As a result of disorders (or diseases) arising from sources outside the brain—as in the case of:

(a) vitamin deficiencies such as pyridoxine dependence;
(b) metabolic disorders such as those which may give rise to low blood sugar or calcium levels, or those in which the body is unable to deal with certain chemical substances derived from food on account of an inherited enzyme deficiency;
(c) toxic states due to ingestion of or exposure to poisonous substances, such as lead, or to an accumulation of breakdown products as a result of liver or kidney disease.

In many individuals, however (perhaps 50% or more), it may not be possible to determine the cause of their epilepsy at any one time. This may be owing to lack of information about the antecedent medical history, or perhaps to the fact that the epilepsy is due to a very slowly growing tumour which may not be detectable for a number of years, or possibly because our present methods of investigation are not sufficiently sensitive.

THE ROLE OF GENETIC FACTORS

It has long been accepted that inheritance may be a factor in the causation in at least some if not all the different varieties of epilepsy. Some believe this to be quite minimal, even negligible, while others, impressed by statistical evidence, have adjudged it to be considerable. Thus, numerous surveys have shown that the incidence of epilepsy among the near relatives of those who suffer from epilepsy may be several times higher than obtains in the general population. This appears to be particularly likely in certain of the primarily generalised epilepsies but less so, if at all, in those of focal (cortical) origin. The situation is, however, complicated by the fact that it appears to be a tendency to epilepsy which is inherited and that there is some other as yet undefined factor which determines whether a person so endowed will actually have overt seizures or not. There seems little doubt that the role of genetic factors is complex and it is still incompletely understood.

THE RECOGNITION OF EPILEPSY

I prefer to speak of the recognition rather than the diagnosis of epilepsy, since the latter term is more properly used in relation to the identification of a particular disease. This practice does, I think, also help to emphasise the fact that the recognition of epilepsy is essentially a clinical matter—it is simply a matter of deciding whether a patient's attacks are epileptic or not on the basis of purely clinical evidence.

The recognition of epilepsy is derived from a history of recurrent, sudden, brief episodes which reflect transient disturbances of brain function which may or may not be associated with disturbances of consciousness. In the first instance, this is given by the patient himself, who is in the unique position of being able to describe, not so much what happens during his attacks, but rather what he experiences at the onset and at their termination. The missing data can usually be supplied by those who have witnessed the episodes and in this way a composite description is obtained, thus allowing a recognisable picture of some kind of epilepsy to emerge. In cases of special difficulty the problem can often only be solved by admitting the patient to hospital for observation of the attacks, which may, if necessary, have to be induced by artificial means.

There are a number of clinical features related to epilepsy in general which are helpful in recognising its occurrence. These are:

1. **Episodes of transient interruption of brain function which come on quite suddenly,** day or night, and without apparent cause. They always tend to have the same pattern and quite often occur in clusters with intermissions of varying duration in between them. Shortly before or just after waking in the morning is a favourite time in some cases and in women there may sometimes be a link with menstrual periods.

2. The recognition of **specific precipitating stimuli** may be decisive, such as a flickering light, a sound, reading, movement, etc.

3. A clearcut description of **tonic and clonic convulsions** associated with blueness of the face, foaming at the mouth, with a sizeable 'hangover' afterwards, leaves no doubt as to the nature of the attack.

4. **Biting of the tongue** — this only occurs in convulsive episodes, of which it is by no means an invariable feature. The biting usually involves one or other side of the tongue in a characteristic manner, such that impressions of the teeth can be seen down the injured margin of the tongue if careful inspection is made soon after an attack. Such a finding does not, to my knowledge, occur in any other kind of attack.

5. **A temporary retrograde amnesia** — this is an inability to recall events which took place several hours before the onset of the attack. Such a memory lapse is recovered later and is characteristic of certain types of temporal lobe epilepsy.

6. **Confused, automatic behaviour** — this may occur during the attack or during the aftermath. Characteristically, it may consist of fumbling with the clothes, looking or wandering round aimlessly, even carrying on with the task in hand. The events of such behaviour cannot be recalled afterwards.

7. **A characteristic aftermath (postepileptic state)**—this is usually only seen after the more severe types of generalised convulsion. It is notable for features such as confused automatic behaviour, headache, nausea or vomiting, mental dullness, aching limbs, soreness of the tongue and drowsiness leading to sleep. Features of this kind are not seen after an uncomplicated ordinary faint which is normally followed by prompt recovery.

INVESTIGATION

The planning of investigations of any case of epilepsy is based upon the findings obtained after taking a full clinical history and the carrying out of a physical examination of the patient. Its purpose is, firstly, to determine the site (or sites) of origin of the seizure discharges, and secondly, wherever possible, to discover the principal underlying cause.

The answer to the first of these two questions is generally sought with the aid of the electro-encephalogram (EEG). The pattern of the attacks may in the first instance afford a guide to their site of origin in the brain; but this is not so if the attacks are always generalised in type and sometimes the pattern of a cortical seizure may be misleading. The value of an EEG examination in this context depends upon how comprehensively it is used and what special methods of activation are used to evoke seizure discharge if it cannot be recorded by purely routine methods. Its use depends upon the fact that in most kinds of epilepsy 'miniature' seizure discharges can be recorded at times when actual seizures are not in evidence. Occasionally a seizure may occur spontaneously during a recording or, if there are special indications, a seizure may be deliberately induced in order that its pattern, site of origin and mode of spread can be closely observed. Unfortunately, under these circumstances so much artefact (extraneous electrical disturbance) is apt to appear in the record that the genuine electrical changes accompanying the seizure are obscured.

Determining the cause of the epilepsy in any given case may often be difficult if not impossible at any particular moment in time. Very often it can only be suspected, or, as in the case of a slow-growing tumour, it may not declare itself for years. A careful scrutiny of the medical history anteceding the first attack may provide convincing or even decisive evidence of a cause and should never be omitted. A plain X-ray examination of the skull can be justified in almost every instance, except perhaps in clearcut examples of light-sensitive seizures and 'classical' spike-wave epilepsy. Such an examination may sometimes yield useful signs, such as anomalies of bone structure and areas of intracranial calcification, and has the merit of being a safe and painless procedure. Brain scanning has been more recently introduced and may be useful in disclosing a tumour or a recently acquired condition, such as a lesion due to cerebrovascular damage. The use of such specialised techniques as contrast radiography involving the injection of contrast media (air, oxygen, dye, etc.) cannot be regarded as a routine procedure in the investigation of cases of epilepsy. Its chief use is when there is a likelihood that the seizures are due to a tumour or a vascular malformation, or if it is thought necessary to measure the extent of organic brain damage.

When there is reason to suspect that the epilepsy may be due to a metabolic disorder the appropriate biochemical tests will have to be undertaken. This appears to be an expanding field which is leading to the uncovering of a progressively increasing number of conditions due to enzyme deficiencies. Appropriate microbiological investigation will only be necessary if there are special indications, such as a history of specific environmental exposure.

Finally, in addition to a detailed inquiry into all relevant psychosocial considerations, increasing use is nowadays being made of the results of a clinical psychological examination. This is often invaluable in disclosing unsuspected areas of cerebral malfunction, in assessing intellectual capacity and in the gauging of attitudes, etc.

GENERAL PRINCIPLES OF TREATMENT

Successful therapy in the widest sense will depend upon the following factors: (*i*) the removal of any cause wherever possible—for example, scars, tumours; (*ii*) the control of seizures by medical means, avoidance of provocative stimuli, etc; (*iii*) the treatment, when necessary, of any associated disabilities, such as dyslexia or a motor disorder; (*iv*) constant attention to social and personality factors.

Removal of Causes

Unfortunately, this is usually only possible in a minority of instances. However, in carefully selected cases an area of damaged brain giving rise to seizure discharge may be successfully removed, provided it is accessible and its removal does not cause unacceptable disability, such as a crippling speech disturbance. The same applies to the removal of benign tumours of the brain, but the fact is that in all surgical removals of this nature there can be no prior guarantee of absolute cure. Even so, the results of surgery can sometimes be very gratifying, to both patient and surgeon alike, and this applies particularly to certain cases of temporal lobe epilepsy. Treatment of underlying metabolic disorders can likewise yield good results so far as control or abolition of the epilepsy is concerned.

Control of Seizures

For most people with epilepsy the best that can be offered is attempted control of their seizures with the aid of anticonvulsant drugs. This kind of treatment can be reasonably effective in some 70% or more of cases. Unhappily this is not always achieved in practice and this

is perhaps more consequent upon inadequacies of application of this method of therapy than upon deficiencies of the drugs in use.

Some 20 different anticonvulsant drugs are currently available, although only about half of them are effective enough to be used as a routine. They require to be carefully selected in any given case and can be administered either alone or in combination. To be effective they must be taken regularly in adequate dosage for relatively long periods of time and a constant watch must be kept for unwanted effects. Although these are rarely dangerous they may sometimes become incapacitating. Not infrequently, therefore, it may be preferable to strike a kind of balance between control of seizures and an absence or minimum of side-effects. Complete suppression of seizures at the cost of drug intoxication can hardly be regarded as successful therapy.

The most commonly used anticonvulsant drugs are applicable as follows:

Petit mal absences (3 c.p.s. spike-wave)—acetazolidine diones (troxidone), succinimides (ethosuximide).

Myoclonic-atonic seizures (Lennox syndrome)—benzodiazepines (diazepam, nitrazepam), carbamazepine.

Generalised tonic-clonic convulsions—phenobarbitone, primidone, phenytoin, sulthiame, pheneturide, carbamazepine, beclamide.

Cortical (focal, partial) seizures—phenobarbitone, primidone, phenytoin. If of temporal lobe origin, sulthiame, pheneturide or carbamazepine may be particularly useful.

It is now increasingly becoming the practice to monitor anticonvulsant therapy by periodic estimation of the blood levels of individual drugs. Reasonably reliable methods are now available and therapeutic levels of the various drugs are being established. With the aid of such a procedure it is possible to ensure that medication is being taken, to detect unexpected fluctuations in blood levels and to identify intoxication and drug interactions.

GENERAL MANAGEMENT AND CONCLUSIONS

There is much to be gained by explaining to patients with epilepsy the nature and implications of their condition in suitable terms. I believe that it helps immeasurably in securing their co-operation in relation to treatment and amply justifies the time spent in undertaking such a task. In addition, they will almost always require advice about education, careers, employment, marriage and so on. They will also often need help with emotional difficulties which may be centred upon family or outside circumstances, or both.

The ultimate aim is to enable the person with epilepsy to lead as normal and as fulfilled a life as possible. This can usually be achieved only through the co-operative endeavours of a team of workers, each an expert in his own particular field, but all with the same purpose in mind.