BRITISH MEDICAL JOURNAL

LONDON SATURDAY JANUARY 24 1953

THE PHENOMENA OF EPILEPSY*

BY

DENIS WILLIAMS, M.D., D.Sc., F.R.C.P.

Physician in charge of Neurology, St. George's Hospital; Physician to Out-Patients, the National Hospital, Queen Square

The question, "Is this epilepsy?" seems to be asked more often and in more various circumstances than it used to be. It may simply relate to attacks of unconsciousness, or to focal disturbance of movement or sensation, but, even so, the answer is unusually important because of its social and economic implications to the patient and his family, quite apart from the problem of treatment and management. Other groups of people are interested too. Employers want to know, sometimes more to safeguard themselves than to protect the patient; public servants have to administer insurance and superannuation regulations; armed Forces have to protect their efficiency from the sudden disruption of a fit or the administrative consequence of its recognition. Teachers want to prevent their pupils from seeing an epileptic attack in another to the ruin of his own schooling; or lovers about to mate calculate the risks of procreation in this respect.

With the assumption of social responsibility by others, and with a more enlightened approach to the study of antisocial and aggressive behaviour, many more people became interested in the answer. Those philanthropists who were concerned about crime and the criminal recognized that some aberrant behaviour or abnormal thoughts might be epileptic, that a cause might be found, and chemical treatment prove successful where psychiatric and social had failed. The child psychologist studying a child with temper tantrums asked whether this might be epileptic. The prison medical officer faced with an intractable Borstal problem, or the psychiatric physician dealing with a long story of intermittent unusual behaviour, has been faced with the same problem. Then lawyers, encouraged by the physicians for the defence in cases of comparatively gainless crime, advance the argument, "This may be epilepsy," so that finally the judge or his jury have to answer the question.

The Diagnosis of Epilepsy

With all this, the ordinary doctor had been seeing many ordinary patients with attacks of loss of memory or consciousness, of muscle-twitching, of feelings of faintness or of unreality, or of transient disturbances of vision, for every unusual one who presented a social problem. Usually there are no physical signs to help him in this diagnosis, for the patient is well when he tells his story, so that he must base his opinion on

minute retrospective facts found by careful questioning of subject and observers. The true interpretation lies in deduction, with the same scrupulous regard for facts as is found in police detection. It is rarely that this method fails, and, allowing the place of experience of the usual and unusual in arriving at a correct diagnosis, the neurologist may succeed simply by taking a more careful history and by sticking to the evidence he has found. In the unusual case, diagnosis may at the time be impossible for want of evidence, or in doubt because of its nature. When this is so, opinions may be given too freely, the term "epilepsy" applied too carelessly; and, when such assumed accuracy seems dishonest, diagnostic uncertainty is expressed as "epileptoid states" or "epileptic equivalents"—phrases which reflect as great confusion of thought in the physician as there is confusion of feeling and behaviour in the patient.

How, then, is the diagnosis of epilepsy to be made? When the distinction is between a faint and a fit there may be no great difficulty, but when the diagnosis rests upon a story of complex disturbances of behaviour, or prolonged disorder of thought or feeling, it may be most bewildering. Clinical observation, aided by electrophysiology and experimental neurosurgery, has very widely expanded the recognized field of disorders caused by epileptic disturbances and has made many definitions worthless. We know that epilepsy can occur without loss of consciousness, without amnesia, without involuntary movement, and without any of the simple features which would enable us to make a certain diagnosis of epilepsy. None of the definitions will do, for they are all too narrow.

The Nature of Epilepsy

Whenever an epileptic disturbance is recognized by the patient or by others there is an abnormal discharge of electrical energy in the brain. It is more accurate to consider the disturbances which are seen in the electroencephalogram as the accompaniment of the disturbance experienced by or seen in the patient rather than as its cause. They are both related to the same cause. Lennox found so much difficulty in defining epilepsy that he called it a cerebral dysrhythmia, having in mind the disordered electrical rhythms invariably present in the attack. In fact, epileptic states are accompanied by a cerebral electrical dysrhythmia, and the electrical disturbances may be closely related to the nature of the clinical attack, but they are not the cause of the attack.

4803

*Address to the Guildford Division of the British Medical Association given on December 11, 1952.

That is not now important; these discharges may occur anywhere in the brain—in the cortex or basal grey matter, in any of its lobes and gyri—and they may have one or a number of patterns of electrical rhythms.

Now all we do, think, or feel depends on our cerebral hemispheres. Simple movements such as a muscle twitch, or more complex ones such as a golf swing or a musical achievement, are mediated through the cells of the motor side of the cerebral integrative mechanism. On the sensory side, where movements are made possible and initiated, such crude sensations as pain or tingling; such complex ones as the awareness of the size, shape, position, and movement of a limb; and such intimate feelings as affection, elation, jealousy, or esteem—all have their physiological origins in cerebral cells. Strangely enough, although the close dependence of the intellectual level on the integrative efficiency of the cerebral hemispheres has been readily accepted, a similar dependence of parallel attributes of personality and behaviour is usually overlooked. This is probably because of the importance put on environmental influences by the dynamic psychiatrists.

It must surely be clear then, if the cortex is so intimately concerned with the whole activity of the organism, that these electrical discharges associated with epilepsy may accompany a disturbance of any movement, feeling, or thought that it is capable of, for they may arise in and spread over any part of the cortex. This cerebral disturbance may be a negative one—the negation of feeling which is unconsciousness—or a positive one—tingling, distorted feeling in a limb, or even an attack of fear or jealousy. Just as the movements of an epileptic attack are a travesty of normal movement because of the crude and unphysiological nature of the epileptic stimulus to the cortex, so the epileptic disturbances of sensation, of thought, or of feeling are caricatures of the normal.

The subject of an uncinate attack experiences an unpleasant indescribable smell that he has difficulty in recalling: unpleasant, because it is a crude complex of smelly sensations; indescribable, because it has not been evoked by a recognized stimulus—it is unnatural: difficult to recall, because it has been caused by an epileptic discharge passing over the olfactory cortex without an extraneous stimulus, and has not followed the integrative pathways within the cerebrum which lead to the storing of impressions, with consequent memory. "It was like nothing on earth, very real at the time, but I can't remember it now," they say. That is in the very nature of the epileptic disturbance. It reflects the ubiquity of the epileptic experience, with the crudeness of its electrical insult to the cortex and its disregard for normal cerebral physiology. We must not forget that, although the epileptic discharge may teach us about the response of the cortex to an abnormal discharge, at most it presents us with a caricature of normal cerebral functions.

What Epilepsy is Not

If what I have said is true, the epileptic attack accompanied by a sudden electrical storm in the cortex cannot have a direct relationship to preceding or succeeding normal events, whether these are in movement, feeling, or thought. The form of the attack is not determined by preceding events; the patient's behaviour after the attack does not flow naturally from it. The attack is an interruption in the flow of activity of the organism. That is why no epileptic attack is "motivated" and why no motivated act is ever epileptic. If intelligent purpose is inherent in an act, the underlying

physiological process must have been that of sensory perception, leading to awareness, central integration of the experience, and then purposive movement. The epileptic discharge does not employ channels of neuronal activity in the brain in such a highly co-ordinated way as this, and it would be surprising if it did so.

Similarly, although epileptic movements or feelings may be fairly complex when the epileptic discharge is disturbing an area of higher motor or sensory integration in the cortex, the nature of the disordered function is related to the abnormal epileptic discharge itself as well as to the normal functions of that area of cortex, so that it is usually primitive and fragmentary. For instance, a discharge in a motor association area may cause such a fragment of behaviour as deviation of the eyes to the left, so that purpose is assumed by the observer in saying, "He looked to the left," or flexion of the index finger, which would suggest that the patient was beckoning. In fact, these incidents were devoid of purpose; they seemed purposeful because they arose in a part of the cortex which normally integrates movement into smooth and purposeful activity. When highly complex thought processes, emotions, or actions are experienced, and particularly when these follow each other in an expected, purposive, or reasonable and natural manner, they probably *are* purposive and in the circumstances reasonable, and not the accompaniment of such a primitive electrical discharge as that which underlies the epileptic state. Epileptic behaviour is always primitive, considered against the natural function of the part of the brain involved.

The epileptic experience is brief, fragmentary, and usually stereotyped. It comes out of a clear sky and fades into it, transient, purposeless, and unfinished. The post-ictal disturbance of consciousness may, however, last for many minutes, and sometimes in status epilepticus the patient may fall from one attack into the next with such brief intervals that it requires care and skill to recognize the individual attacks, but this can usually be done. Any one epileptic attack lasts for about the same time as the electrical discharge in the electroencephalogram, and, although all brief attacks are not epileptic, no continuous prolonged disturbances of behaviour, thought, or feeling are entirely epileptic in origin. I say entirely, because a patient may hit his head in an attack and so be confused for some time after, have another attack, or develop a hysterical disorder during the post-ictal period of lowered consciousness. The epileptic disturbance is in itself brief.

Lastly, because of its nature the epileptic state is episodic, and indeed one of the unsolved problems of the epileptic discharge is why it ceases, why it appears to be self-limiting. The short stereotyped episodes are usually recognized because they stand out in clear contrast to the intervening normal behaviour.

It should now be clear that no positive definition can be applied to epilepsy, because the epileptic disorder may interrupt or mimic any aspect of normal movement, feeling, or thought. The content of an epileptic attack cannot be continuously related to preceding or subsequent normal events, it cannot be demonstrably and intelligently purposive, and it is not usually prolonged. The phenomena of epilepsy tend to be episodic, stereotyped, primitive, fragmentary, and unnatural—at variance with usual experience.

Faints and Fits

In a discussion at the Royal Society of Medicine recently it became clear that at times it is impossible to make a sharp division between epilepsy and syncope, although the distinction can nearly always be made for any one attack upon clinical grounds alone. It is probably best to reserve the word "epilepsy" for those cases in which the loss of consciousness is *primarily* due to a cerebral disturbance, and "syncope" for those in which it results *primarily* from a fall in blood pressure. The important qualification here is the word "primarily," for loss of consciousness from either cause may follow disorders of many kinds beyond the range of the present discussion. Whether the *secondary* effect of

the disorder is epileptic or syncopal, the chain of events is in each case relatively constant. The major fit seen in constitutional epilepsy or that symptomatic of bodily disorder is the same in its pattern. Indeed, the nature of the epileptic attack seems to depend more upon the part of the brain affected and the particular brain being affected than upon the causal agent. I have seen volunteers subjected to oxygen intoxication in standard conditions, most of whom had a convulsion at a critical level, but some of whom had an attack of aggressive behaviour—so-called psychomotor epilepsy. The same is true of fainting: the actual sequence of events in the faint is similar whatever the cause.

Syncope may be distinguished from epilepsy often by a cause—postural, emotional, or physical; a fairly long warning, with the characteristic experience, "I feel faint"; protective actions such as sitting; and early onset of pallor, with hot or cold sweating. Consciousness usually returns without any confusion or sequels apart from sweating. The attacks are infrequent. The cerebral anaemia of syncope may lead to a convulsion, but then the primary diagnosis is syncope. Sometimes the two conditions may be very intimately linked, as in the Stokes-Adams attack, in which rapid fall of blood pressure caused by cardiac arrest produces convulsions through cerebral anaemia.

Syncope simulating epilepsy was illustrated by the following case.

A man of 23 had a history of three convulsions. He had had six attacks of loss of consciousness in his life: the first when standing before the blackboard in school; the next after a hearty meal, exhausted from a day's climbing; the next, seeing a crushed thumb dressed; one during a febrile illness with diarrhoea; and two others in tropical weather when standing. In the last three he had had a convulsion, and he said he had been unable to sit down in time to prevent loss of consciousness. He had always suffered from feelings of faintness on rising quickly. He was 6 ft. 2 in. (188 cm.) and only 10½ st. (66.7 kg.), with a pulse rate of 50 to 60, and a diastolic pressure of 55 mm. Hg. The electroencephalogram was normal.

Here, then, was repeated syncope complicated by convulsions, the diagnosis of epilepsy being unjustified. On the other hand there is this case:

A man aged 35 had for six months had attacks of faintness, which had gradually changed their character. They began with fleeting feelings of unreality; then these were accompanied by pallor and goose-skin, later by sweating and the sensation "I feel faint," and finally by a sense of familiarity. Later he lost consciousness in these attacks, which were the result of a malignant glioma arising in the anterior part of his right temporal lobe and invading the lower part of his frontal lobe. This was therefore an example of focal epileptic discharges spreading to a part of the cortex known to bring about modifications in cardiovascular activity, and in consequence causing syncopal changes as part of the epileptic disturbance.

The value of unusual cases of this sort is to emphasize the importance of recognizing the nature of the primary cause of the attack and of basing the diagnosis upon that rather than upon the superficial similarity of the ultimate attack to a fit or a faint.

Forms of Epilepsy

Whatever the cause of the attacks and whatever the electroencephalogram may show, the initial division of the epilepsies is bound to be based on the nature of the observed fit. It so happens that such a division of clinical types of attack is the most useful and fundamental of any, as I hope to show later. It used to be enough to recognize epilepsy, with a division into its major and minor forms, but now more detailed division is essential to the most successful management and treatment of the condition. For instance, minute disturbances of consciousness (*petit mal*), which are superficially similar, may be caused by slight attacks of each of the different forms of epilepsy, and only most careful observation or questioning may distinguish them from each other.

"Absences"

The French word *absences* immediately calls to mind the frequent *petit-mal* attacks of childhood. Here there is a

brief loss of consciousness, without involuntary movement, sometimes with loss of muscle tone, causing the subject to drop what he is holding, or to fall and as quickly pick himself up. The features of the attack are loss of consciousness and loss of muscle tone. There may be hundreds of attacks a day; they are usually worse in the mornings, and they tend to get less frequent as life advances. They are invariably accompanied by spike-and-wave complexes in the electroencephalogram. These attacks (*juvenile petit mal*) are accentuated by alkalosis and hypoglycaemia, are not improved by the hydantoins, and rarely improved by sedatives, but respond favourably to the diones and sometimes to such stimulants as caffeine and the amphetamine group of substances.

They may be closely mimicked by other epileptic attacks having a different clinical significance and a different pharmacopoeial response. Very brief clonic attacks with almost imperceptible twitching round an angle of the mouth or in one limb may cause interruption of attention, thought, or consciousness similar to that in *juvenile petit mal*. Tiny focal sensory attacks may be associated with a similar change; or frontal adverse attacks, in which the head and eyes turn towards one side, may give a similar momentary amnesia. Any evidence of associated disturbance in a patient with brief amnesic attacks should be assessed carefully—muscle-twitching, movements of limbs or eyes, licking, smacking of the lips, a local change in sensation, a fleeting smell or taste, a change in mood, or disturbance of time or reality. Any one of these may lead to the realization that the brief attacks are not *juvenile petit mal*, with consequent differences in investigations, management, diagnosis, and treatment.

Circumstances in which search for features such as these may be profitable are when the attacks begin after puberty and especially in adult life, when they follow a cause such as a head injury or cerebral disease, when they are infrequent (less than half a dozen a day), when they are less frequent early in the day, or when a number herald the onset of a bigger attack. In these circumstances we may correctly predict that the electroencephalogram will not show the wave-and-spike discharge, and in its absence during a clinical attack the diagnosis of *juvenile petit mal* may be excluded.

The following are two examples of *petit mal*—not classical or *juvenile petit mal*—representing symptomatic epilepsy, and consequently failing to respond to tridione or paraldione.

A child aged 13 had had attacks for four years. She might have two in a day or six in a week, but would sometimes be free for two weeks. They occurred at any time of the day. They were stereotyped, and in them she would stop what she was saying or doing, look vacant, mumble for a few moments, go pale, and then resume her activities. She had had meningitis when aged 3 and was the only left-handed member of her family. This, then, was symptomatic epilepsy with transient "purposive" behaviour in speaking—probably a left Sylvian lesion. The electroencephalogram showed abnormality, made up of medium-voltage two- and three-a-second waves with focal spikes in the anterior part of the left temporal lobe. Her attacks had been made worse with tridione.

The second example of symptomatic epilepsy with brief attacks simulating *juvenile petit mal* was in a 30-year-old woman who for six months had had momentary attacks in which she was amnesic and out of touch with her environment, and in which it transpired that she looked to the left side and sometimes turned her head to that side too. At first she had one every week or two, but they then increased to one or two a day. They were caused by a right frontal glioma which showed as focal slow waves without any "epileptic" discharges in the electroencephalogram.

Attacks with Clonus

Attacks of clonic muscle-jerking have many features in common, whether they are localized to a digit or a limb or involve the whole body. When both sides of the body are involved in a motor fit consciousness is lost except in rare instances. The spasmodic contraction of muscles without regard to their functional relationship that occurs in these attacks is accompanied by very fast waves (spikes) in the electroencephalogram. These may be focal or generalized,

single, in transient bursts, or more prolonged as in a major convulsion. The major convulsion is in fact the ultimate intensity of this form of epileptic discharge. Focal clonus probably indicates a local organic lesion, and when it begins in adult life the possibility should be considered of the lesion being recently acquired. If the attacks change their character progressively the lesion is probably an advancing one. But the pathological nature of the lesion does not alter the fundamental character of the clonic attack. After the causal lesion has been investigated, and if possible recognized, chemical treatment of all attacks with clonus follows the same plan—usually the use of barbiturates and hydantoins.

Attacks with Co-ordinated Movements

There are many attacks in which the movements are smooth and natural, not clonic. Here the epileptic disturbance has arisen in a part of the cortex responsible for co-ordinating and integrating movements. Because of this the cortical discharge causes response in the body which is physiologically integrated, so that, for instance, when muscles contract there is inhibition of antagonists. This is in contrast to the unphysiological simultaneous contraction of agonists and antagonists in the tonic-clonic fit. Because the movement of limbs in these attacks is necessarily co-ordinated, it appears to the observer to be reasonable and to have purpose, although as it is invoked by a primitive epileptic disturbance it is devoid of purpose. Attacks of this sort have been called "psychomotor" or "epileptic equivalents." They are in fact focal epileptic attacks involving cortical association areas, which used to be called silent areas, but which are responsible for the integration of feeling and movement. The movements or feelings which are invoked are consequently more elaborate than in other sorts of attacks. They are commonly seen when a focal fit begins in a temporal or a frontal lobe.

The movements which are seen in these attacks may range from a single act, such as deviation of the eyes, licking, swallowing, or movement of a hand, but they include complicated rituals, salaams, and simple aggressive acts. Most of the prolonged purposive movements seen in association with epilepsy occur in the post-epileptic confusional state, are post-ictal, and are not strictly epileptic.

Post-ictal Activity

It may be difficult to tell when the abnormal behaviour is occurring during the actual epileptic fit (ictal) or after it (post-ictal), particularly when an insignificant and brief fit is followed by a long post-ictal confusional state causing the restless behaviour of a fugue. As a rule the behaviour seen in the fit is apparently spontaneous, and, being the result of the epileptic disturbance of the brain, is stereotyped, short-lived, and without relation to circumstance. The post-ictal activity, whether it is restlessness, aggression, or a fugue, is due to the state of confusion, and so it is simple in its form, evoked by stimulation; when purposeful, primitive, or stupid in its purpose its form is often determined by past experience. It seems automatic. The confused person just walks, or fumbles with clothes, undresses routinely or blindly, resists help or interference. While there can be no real purpose in the ictal activity, any purpose in the post-ictal is immediate and very crude. No prolonged, elaborate, and intelligently purposeful fugue state is epileptic. Very rarely the actual epileptic attack may consist of a lowering of consciousness (confusion) and so simulate the post-epileptic state described in this paragraph.

Sensory Attacks

As sensation must be experienced by the patient there cannot be a sensory equivalent of the major tonic-clonic fit, since consciousness cannot be retained when the epileptic discharge has spread over both hemispheres. The form of sensory disturbance in epilepsy may be negative (loss of sensation) or positive (a hallucination). These may affect common sensation, special senses, or emotions, and mixed forms are common.

The commonest kind of sensory disturbance encountered in epilepsy is very crude—simply tingling at an extremity; but disturbance of proprioception with changes in the shape or position of a limb are met. Epileptic pain is uncommon, but is experienced as a mixture of intense pains, often with a sensation that the limb is in an unnatural position. While tingling is the sensory counterpart of a focal clonic fit, the more complex disorders of feeling, relating to the shape of a limb and causing sensations of bodily or spatial distortion, are the counterpart of the attacks in which co-ordinated movements occur.

Similarly, although when special senses are involved in the epileptic experience they are felt in a most primitive manner—as an unrecognized smell or taste, or flashes of white light—they may be co-ordinated with elaborate formed hallucinations when the epileptic discharge happens to involve one of the special sensory association areas. The most common examples of these are the elaborate, animated, and coloured scenes witnessed by the subjects of lesions in the parieto-occipital area. When even more highly organized areas of cortical integration in the temporal lobes are affected, hallucinations made up of the blending of several sensations may be indivisibly experienced. These may be accompanied by spontaneous and transient emotional disturbances.

The episodic disorders of emotion experienced during temporal lobe attacks have not yet been studied in sufficient detail, perhaps because they are easily overlooked by the questioner, but also because they may suggest to him a psychological disorder rather than a physical disturbance. They may contain any possible emotional experience, any combination of emotions, or any linking of emotion with simpler sensations, and they may evoke appropriate visceral or somatic behaviour. They must be recognized as being epileptic if the patient's problem is to be fully understood.

Prolonged Disorders of Behaviour or Feeling

In status epilepticus, focal or general, abnormal states of movement, sensation, or consciousness may last for many hours. It is usually possible to recognize the individual attacks of the status and so to establish the responsibility of epilepsy for such a prolonged disorder. Otherwise some other cause for the disorder should be sought, since a single epileptic attack is so short-lived. Prolonged amnesic states, fugues, stupor, coma, or aggressive or unsocial behaviour are unlikely to be epileptic in the absence of status, for even post-ictal confusion lasts for minutes rather than hours. In particular, as I have said, activity which is motivated, or which uses past learning, cannot be epileptic. The soldier, absent without leave, who finds his way home by road and rail with no memory of the process, could not have done so in an epileptic attack; neither can the criminal who has amnesia for an act of violence directed against a selected subject with an appropriate weapon, however obscure the motive or violent the act. In cases of this sort organic causes having been studied, natural or neurotic motives must be sought, and any evidences of a psychopathic or psychotic personality should be assessed.

It might be said in criticism of this didactic and rigid distinction between epilepsy and other states that the borderland between epilepsy and psychopathy is a narrow one, hardly explored, and that the basis of neither state is understood. That is no justification for a mongrel diagnosis such as that of "epileptoid state" or for the subterfuge of calling inexplicable behaviour epileptic upon tenuous or inappropriate evidence.

A sixth revision of the *Index of Modern Remedies* has been published by the *Scottish Chemist*, 240, Albert Drive, Glasgow, S.1. It includes much information on proprietary preparations and takes note of the 1951 addendum to the *B.P.*, the *National Formulary*, 1952, and the *Pharmacopoea Internationalis*. It is a most useful handbook for all who need an up-to-date guide to modern drugs. The price is 5s., and in London it may be obtained from H. K. Lewis, 136, Gower Street, W.C.1.