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CONTENTS

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Epilepsy and its Variations*

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Epilepsy is an extremely common disease, although actual statistics on racial and geographical distribution are virtually unobtainable. Turner in 1907 estimated that the ratio of epilepsy per thousand of the population in England ranged between 2.9 and 0.4. In the last nine months I have seen 57 cases of epilepsy, and these form the basis of this report. Of these, 54 were Europeans, two Indians and the other was Coloured. I have not included any African in this report, for although it is a common disease amongst them, I have seen insufficient cases myself. Of these cases, 33 came from Salisbury. If the European, Indian and Coloured population of Salisbury is about 80,000, this gives a personal incidence of 0.4 per thousand. Now it is clear that only those cases have been referred for consultation which are either new or are giving trouble, and if as many as one case in four were referred, this would give an incidence of 1.6 per thousand; in fact, I doubt whether as great a percentage of these have been referred, and therefore the incidence must at any rate be higher than that.

Unfortunately epilepsy has a very unjustified reputation in the minds of the lay public and one has to be very careful about mentioning it as a diagnosis. The name brings a gasp of horror to most parents and a wave of revulsion amongst friends. V.D. is certainly more respectable than epilepsy. It conjures up the spectre of the victim uttering a loud shriek, to fall writhing and frothing on to the path; then, when the fit has finally passed, he is expected to get up in a state of post-epileptic automatism and stalk off to commit yet another brutal murder. It is unfortunate that this rare aspect of epilepsy has been so played up in the press and in fiction that this is now fixed firmly in the minds of the majority as the typical feature of elipepsy. In fact, of course, not one epileptic in 10,000 follows this pattern.

The word epilepsy means a seizure, and Hippocrates used it to suggest that the disease seized the patient and prevented the exercise of his own free will. Definitions have been many and varied and none is entirely satisfactory. It consists of an uncontrollable and abnormally stimulated burst of nervous energy manifesting itself by temporary unco-ordinated and pointless activity, and frequently accompanied by loss of consciousness.

In olden times the cause was completely unknown, and frequently the disease was referred to as "Morbus Sacer," or the Sacred Disease. With the advance of medical science it has become apparent that some cases of epilepsy have in fact a relation to organic disease, and hence the disease has been divided into two groups—symptomatic and idiopathic. At first the symptomatic group was extremely small and the majority of cases fell into the idiopathic group. However, as more diagnostic tests have been introduced and our understanding of the disease has increased, we are slowly whittling down this group of apparently self-productive seizures and increasing the number that fall within the symptomatic group. In fact, so far has this process gone on that I think it is fair to say that we have now recognised that all epilepsy is symptomatic and that sometimes we know the underlying disease and sometimes we do not. The consequence is that I feel that the term "idiopathic" should be abandoned altogether, and if epilepsy is to be classed in this way it should be classed as symptomatic and "cryptogenic" or of hidden cause. Of course, although we are one stage further on in realising that there is an underlying disease in each case of which epilepsy is a symptom, the actual mode of production of the seizure itself still remains a mystery. Only when more work has been done on cellular metabolism and neurone irritation will the problem of how irritants produce seizures be solved.

There is immense confusion concerning the various terms that are used to describe this condition, though epilepsy, convulsion, fits, seizure and grand mal are used more or less synonymously without much regard to their true meaning, and with the term "petit mal" tagging along behind. The entire problem is related to the history of the disease. In the first instance, and even up to the present time, the pre-occupation of the physician has been with the motor element of the attack and with the loss of consciousness—the two visible phenomena. Even to-day fits are described as commencing only at the point when motor symptoms make their appearance, and quite regardless of whatever subjective sensory symptoms that the patient in the meanwhile may have experienced, so that warnings or aura and prodromata, although described, have been excluded from

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the content of the fit itself. Even until very recently only two types of epilepsy have been recognised—grand mal or major convulsions associated with unconsciousness, and petit mal or transient unconsciousness unassociated with motor movements. Both these are virtually outmoded, since they described epilepsy in terms of a disease, and certainly the term grand mal should be abandoned. The term petit mal may be retained under certain circumstances, and we will discuss this later.

A tremendous variation even in the motor content of the seizure has been described, let alone a consideration of the other types, so that it is no longer possible to use only two terms, grand mal and petit mal, to describe the many and various fits that occur. The words seizure and fit may be used synonymously, since neither of these suggests any motor element, while convulsion means that during the course of the attack there was some motor element.

The terms general and focal have been used in relation to motor convulsions. On the one hand the term generalised convulsion is used to denote loss of consciousness and generalised motor activity, while in a focal convulsion one or several parts of the body are involved; there may or may not be a loss of consciousness, and the remaining parts of the motor system may be unaffected. It is now becoming clear, however, that virtually any area of the brain may give rise to this abnormal activity; that depending on the strength of the abnormality and the susceptibility of the rest of the brain to it, depends the spread to other parts and their involvement by this abnormal process in a generalised or focal seizure. Thus a focal seizure need have no motor element and only some part of the sensory system or of the affect may be involved. Seizures clinically, therefore, may take a wide variety of forms, both in their mode of presentation and symptomatology. I am therefore beginning to feel that any seizure which begins with a clear-cut aura, although it proceeds to a generalised convulsion, should be called a focal attack.

On the other hand, from the standpoint of the electroencephalogram, a seizure may show much the same sort of activity from any area of the brain which is involved in this epileptic activity. It is therefore important for us to realise that symptomatology and type of seizure are in many cases no more than a brief clinical localisation of the irritated area, and that from the basic neurophysiological standpoint a seizure starting in one area is virtually identical with that starting elsewhere.

All these terms were introduced before the electroencephalogram enabled us to study the mode of action of the brain electrically, and much of the difficulty existing to-day is due to the fact that we are trying to use terms which were introduced to describe motor effects only and carry them over into neurophysiology, and use them to describe the findings of electroencephalography.

In the past all attention has been focused on the motor side and this has been minutely described, but the aura and prodromes have been dismissed in but a few words. The fact that most seizures have a motor element simply means that most seizures are sufficiently severe to involve the motor area, although it is possible that this area is more susceptible to epileptic activity than other parts of the brain.

I visualise a seizure as the boiling over of the pot or the eruption of a volcano, and I believe that undue attention has been focused on the motor element, so that the true perspective of the whole has been lost and the importance of the prodrome and aura and post-seizural state have been under-estimated.

A seizure should be described as consisting of (a) prodrome; (b) aura; (c) the main phase; (d) the post-seizural state, and the whole must be seen as blending together to form a continuing sequence in which it is incorrect to separate any one part as being the fit. The fact that the motor side has been stressed to the exclusion of not only the prodrome and the aura, but also all other types of seizure, has given an incorrect slant to the whole picture.

The prodrome is the first sign that the pot is beginning to boil. It is the little bubbles that come up from the bottom and break the surface and show that something is going on underneath. Prodromes may take all sorts of forms. Nine patients in this series can tell well in advance when they are going to have an attack. Headache is the commonest of these symptoms and three patients suffered from it. On the other hand, two others become intolerable and very crotchety, while one patient feels extremely well before his attacks. Nausea and constipation and malaise also are experienced.

As the seizure approaches, the prodrome shades off into the aura. Aura means a breeze, odour or gleam of light and is used to describe the peculiar sensations felt by the patient immediately preceding the attack. Frequently it is difficult to tell where prodrome ends and aura begins. The aura is the milk boiling in the pot as it starts to rise up the side, and

sometimes by a tremendous effort of will the patient may be able to snatch the pot from the stove, as it were, and break the seizure at this point. More often than not, however, this proceeds relentlessly into the main and involuntary phase of the attack.

This involuntary phase is of course incorrectly known as the "fit," because in the motor convulsions, first described as grand mal, this was the really visible component. It may, however, take any form, depending upon the part of the brain involved. Table 1 shows the type of involuntary activity found in these 57 cases.

Table 1

TYPE OF SEIZURE IN 57 CASES

Generalised convulsions	25
Amytonic	11
Focal motor	12
Petit mal	1
Headache	3
Temporal lobe	1
Breath-holding, etc.	1
Funny seizures—trembling, swallowing, ears go red, etc.	1
Visceral	1
Affective seizures (laughter)	1

Although the generalised motor convulsion is the most common, it will be seen that many other parts of the brain may be affected on a localised basis only.

Generalised motor convulsions are often described as having a tonic and clonic phase. First of all the patient goes stiff and remains in this state for some time, all limbs being affected. The limbs start to jerk, and rather than call this the clonic phase, I am not sure that it should not be called the intermittent tonic phase—that is, if it has to be differentiated from the first portion of the motor element at all. It seems to me that it really represents a dying away of the seizure from the motor area.

As the main stage of the seizure, whether it be motor, sensory or other, passes away, the patient may fall asleep or else wake up feeling perfectly normal. Sixteen patients fall asleep or are tired for varying periods after the attack, especially after the generalised convulsions, while others complain of confusion or headache. The remainder soon become normal.

The typical seizure then may be seen to consist of a little bubbling at the pot, a sudden warning as the flood tides engulf the patient, the involuntary crescendo and the finale as the seizure passes off and the patient returns to his normal interseizure state.

Not all seizures in any one patient may be the same. A patient may have both the major convulsions as well as minor fleeting vacancies (nine patients), which most of the time I would think are sub-clinical or aborted attacks. These are often called petit mal, but this is incorrect. Petit mal is the name given to a particular type of vacant attack, lasting only a few seconds and invariably characterised by three per second spike and wave abnormalities in the electroencephalogram, and because this has been accepted as a clinical entity the name will remain likewise where grand mal must be rejected.

It is perfectly true that these aborted seizures sometimes do not have the same aura as the typical major attacks from which the patient may suffer, and it is possible that a patient may suffer from two different types of seizure. However, in my opinion this is unlikely in most cases in this series. Whereas there are a number of precipitating factors such as anxiety, family disharmony and tiredness, these are only predisposing in a general way, while repeated flashes of light are very epileptogenic and can be used to drive an abnormality and precipitate a seizure. There are, on the other hand, certain attacks which may be almost totally reflex and require a specific stimulus to set them off.

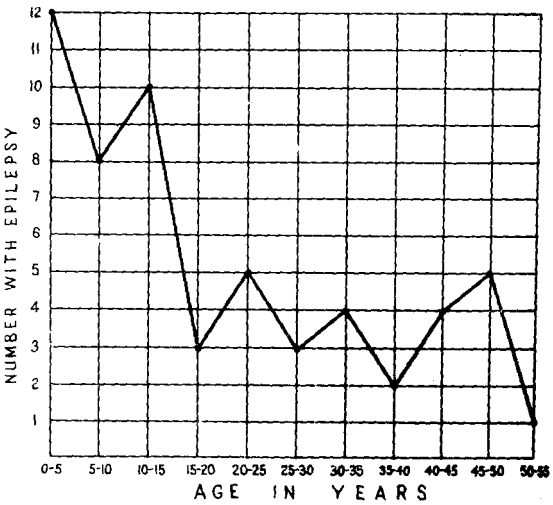

Table II—Graph giving the number of epileptics at different ages.

The ages of the 57 epileptic subjects in this series range from a few weeks to 51 years at the time of occurrence. It will be seen from the incidence table that apart from those occurring in infancy, that there are insufficient to construct any sort of an age curve. In any case, an age curve would be of no significance unless correlated with the underlying disorders, but it is interesting that there are no old epileptics.

Every case of epilepsy should be thoroughly investigated to try and elucidate the underlying pathology. It was not possible for every case to have an electroencephalogram, as this facility is not yet in existence in Salisbury. Whenever a seizure had shown any form of focalisation, however, I believe that definitive tests such as cerebral arteriography and pneumoencephalography should be performed. Where there is a strong familial tendency these tests are not quite so important. Certainly any cases of convulsions appearing in a person over 30 should, in my opinion, be considered a brain tumour until proved otherwise. Only 11 patients have been submitted to electroencephalography, 24 have had pneumoencephalograms, 20 have had arteriography. In some patients these three tests overlap. Otherwise investigation has been by general examination, lumbar puncture, skull X-ray, etc. The diagnosis in these 57 cases is as follows:—

Table 3
DIAGNOSIS IN 57 CASES

Cryptogenic	35
Arteriosclerosis and atrophic	3
Angioma	2
Brain tumour suspect	2
Post-infectious	8
Brain tumour	1
Febrile	1
Microcephaly	2
Post-traumatic	3

On physical examination only 16 patients showed any neurological abnormality. Neurological changes are particularly marked in the very young children, where, in nine children whose seizures started below the age of 15 months, five showed abnormality, varying from hemiplegia to severe personality difficulty.

It will be seen the commonest is one or more seizures per day, but in most cases these were of the minor type unassociated with unconsciousness. Major seizures tended to occur more infrequently and presumably have a longer "refactory period," if I may use that term.

Table 4
FREQUENCY OF ATTACKS

One or more per day	13
One or more per week	8
One or more per month	7
One or more in three months	4
Six months	1
Annually	3
Associated with fever	1
Unknown	9
Seen following first attack	11

There seems to be a certain routine of time about some seizures. The vast majority may occur at any time of day or night, but some occur at specific points in time, either on waking (one case), or on falling asleep (one case), or during the night only (three cases), or shortly after waking—in fact, while shaving (one case). I do not know what the significance of this is, but presumably it must be related in some way to the fact that at those moments the cerebral defences are down and the seizure is able to take charge. The seizures do not seem to differ in any way from those which are liable to occur at any time.

Twenty-four patients stated that they were aware of an aura, but there is no saying that others do not have one also. Unless they complain of this to those near by at the time of the attack, a post-epileptic amnesia may cause them to forget it.

Miss H., a 41-year-old school teacher, was referred by Dr. Fleming, of Gatooma. She had an intracranial operation for cerebral aneurysm one year ago. She is now getting black-outs, without any aura that she recalls. The other day, when she suffered a black-out in her car, it was found afterwards that the engine had been switched off and the hand brake applied, so that clearly she had some warning, although she does not recollect it.

Frequently patients complain of a visceral aura, which may consist of a tight feeling in the stomach, nausea or dryness of the mouth. The insula is known to control gastric movements, and irritation of this region is known to be preceded by such an aura. Olfactory or auditory aura may be related to the temporal lobe, but visual aura do not seem to be quite so regularly related to the occipital area, as

Table 5

TYPE OF AURA

Visual	7
Movement some part	3
Visceral	3
Paraesthesia	2
Blank period	2
Giddiness	2
Cry	1
Sleepy	1
Headache	1
Unknown	1

far as I know. Nonetheless it seems to me that the aura can be very important in localising the source of the abnormality. It is of course by no means as important as the electroencephalogram, but it helps immensely, not only in the examination of the seizure and its treatment by surgical means, but also from the anatomical standpoint of cortical localisation.

We have seen earlier how the generalised convulsion is by far the most common type of seizure (25 out of 57). Only nine of these 25 complain of an aura, as do 7 out of 12 focals and 5 out of 11 amyotonics, and this means that 27 of these patients alone are liable to have periods of unconsciousness without any warning, except possibly some mild prodromata. This underlines the tremendous importance of controlling seizures completely by any means available.

All the patients who suffered from either generalised or focal motor seizures suffered also from unconsciousness. Seizures unassociated with gross motor involvement, petit mal, affective, visual, trembling, etc., are associated with unconsciousness in 12 out of 20 cases.

The typical generalised convulsion which has been called grand mal consists of a prodrome, aura, motor phase, tonic, clonic and then post-epileptic state, and these seizures may result from a focus anywhere within the brain or from many foci within the brain. The patient may at the same time be incontinent and bite his tongue. These seizures are known to everyone and I do not propose to spend any more time on them. Much more interesting are seizures affecting local parts of the brain and not spreading throughout it.

(1) Focal motor seizures are presumably seizures resulting from a localised area of irritation and, depending on the degree of irritation and the anticonvulsant activity of the brain, hangs the extent to which the seizure spreads to other parts of the brain.

(2) An interesting type of focal seizure is that which involves the temporal lobe, producing what is known as psychomotor epilepsy.

K.C., a young man aged 21, was referred by Dr. Robert Fynn. He had a black-out in January, 1957, when he fell down and remembers nothing, but he had a convulsion in which the movements were apparently purposive. Since that time he has had numerous attacks in which he has not been completely unconscious and others in which he has apparently just fallen asleep with a sort of far-away feeling. I believe that this man has epilepsy originating in the temporal lobe, and although we have not done an electroencephalographic tracing, owing to parental resistance, I believe that this would show an abnormality in that area. Dreamy states, the deja vu' phenomenon, memories and so on are very common in temporal lobe epilepsy.

(3) Another is one involving the affect and is unusual but interesting.

Mr. D.M., aged 41, was referred by Dr. Gilbert, of Mabelreign. He stated that since August of last year he had had bursts of uncontrolled laughter, totally unrelated to anything that was going on at the time. He had some trembling of the muscles on exertion. Physical examination was not very remarkable, but electroencephalography showed a mild diffuse abnormality and an arteriogram showed tortuous and sclerotic vessels. Epanutin has diminished though not completely abolished the attacks. I recall a similar case recorded in the literature, but its reference I cannot find, in which a student would suddenly be compelled to stand and make a speech for a short while. He always said the same thing, and then would sit down looking rather dazed. These I take to be similar cases involving the personality, and in some way may be a sort of frontal lobe epilepsy.

(4) Headache frequently forms a component of the prodrome or aura; occasionally, however, it may be present as the uncontrollable phase, and under these circumstances must be considered an epileptic variant involving whichever particular region of the brain is productive of headache. The headache may last one or more hours and is resistant to all the usual anti-cephalgic medication. Electroencephalography may show localised or generalised abnormality and anticonvulsant therapy may prevent the headaches from coming.

Mr. J.T., aged 44, was referred by Dr. I. Zinn. He gave a history of headaches lasting about two hours and coming on once per week. The headaches were diffuse, although very severe, and when he had them he had to lie down. They were preceded by an aura of a dry mouth and a feeling of thirst. He had been tried on all the usual headache medication, including the anti-migraine drugs, but without any result. Electroencephalography had not been performed, but he was placed on gr. ½ luminal three times a day, since when he has had virtually no headache and feels immensely

better. It is possible that it may be said that the luminal merely acts as a sedative and for this reason makes him feel better. However, I am not prepared to concede that point.

A second case is that of Miss S.B., an 18-year-old student, who was referred by Dr. A. C. Cairns. She gave a history of eight years of right side headache, with numbness on the left hand. Recently this has spread to the foot. The attacks always come on during the day and last several hours and are always heralded by a flicker across the eyes, especially on the left side, and difficulty in focussing. Towards the end of the attack, which lasts several hours or a day, the patient vomits and then falls asleep. They were always aggravated by worry and tiredness. All the usual drugs against migraine had proved unavailing and she was put on 1½ gr. of Epanutin three times a day. Since that time she has had a small number of attacks, and a number of emotional situations which might have been expected to precipitate one have not done so. Electroencephalography performed by Dr. Ellerker is normal. Here again it is possible to claim that this disease is not epilepsy but migraine. However, the patient does not respond to any therapy for migraine, and the fact that she has had one normal interseizure record is insufficient to prove that epilepsy is not present. Unfortunately we have not at this time managed to get a tracing during a headache, and since she is so much better it is unlikely at this time that we shall. Epanutin is not a strong tranquillising drug: in fact, much less so than luminal. Here again I am convinced that this is much more of an epileptic attack than it is of a migranous one. The whole relation of migraine to epilepsy is not clearly understood and certainly there are a great many headaches which shade between the two, and it is possible to lay claim that since migraine has an aura and a main phase of headche, all cases should be included as a special form of epilepsy.

(5) An interesting and unclassifiable seizure occurs in a small boy (B.P.), aged 2¼, who was referred by Drs. M. S. Barnard and G. Brander. The child suffered from meningitis at nine months, with paralysis of the right side and twitching of the left side, from both of which he recovered. Since that time he gets attacks about four times daily in which his face becomes red, he trembles and swallows, and he runs to his mother and hides in her skirts. The whole thing lasts approximately two minutes. Pneumoencephalography showed subdural fluid and a bilateral trepine allowed a small quantity of this to escape. He was put on ¼ gr. luminal three times a day, since when he has been virtually free of attacks. This is a case where seizures follow meningoencephalitis, but they are certainly of a very peculiar nature and their source and mode of occurrence are a mystery—possibly they arise near the third ventricle.

Some other types of seizure are worth mentioning.

(6) Myoclonic jerks occur as sudden jerking spasm, usually involving all muscles. These jerks may be made more or less continuously or only occasionally and intermittently, and there must be some form of bilateral motor seizure.

(7) *Epilepsia partialis continua* consists of continuous twitchings of one part of the body —for example, the thumb—which periodically bursts out into a major convulsion. This is just status epilepticus, which alters its intensity from time to time, sometimes giving major seizures, sometimes giving only minor ones.

(8) Narcolepsy or attacks of sleepiness must be focal epileptic attacks involving the sleep centre.

(9) A condition known as "carotid sinus epilepsy" has been described in which attacks of unconsciousness unassociated with change in blood pressure or pulse can be produced by stimulation of the carotid sinus by rubbing it with the thumb. I have never seen this condition, although it is well known, but here is a case which appears to be reflex in origin.

J.N. is a 12-year-old cerebral palsied child, referred by Dr. Hutchinson, of Gwelo. She suffered from encephalitis at 15 months. Since that time she has had difficulty in walking and making well-controlled movements and is severely mentally retarded. She also suffers from rather peculiar seizures. They always occur at night, and it appears that she tries to wake up and get out of bed. I suppose because her bladder is full and she desires to empty it. On the way while she is doing this she develops a seizure and becomes staring, still and rigid, her arms rise up and her stomach is distended, and she makes a croaking noise. She may or may not lose consciousness. Her mother in the next room immediately comes in and massages her stomach, when she brings up wind and apparently wakes up. She then falls asleep.

This is some kind of peculiar visceral seizure, either associated merely with the act of waking up or else with some reflex which provides the trigger mechanism. It seems to me unlikely that this is due merely to the act of waking up, since the patient wakes up every morning without having an attack and these only occur during the night. The mother presumes that the child is getting up to try and get her chamber pot, and it seems that the visceral reflex that is set off at that time is in some way the trigger mechanism for the whole seizure. How these seizures come about is not known.

Here I would like briefly to mention the concept of epilepsy of the spinal cord. Muscle spasms owing to disseminated sclerosis or following trauma to the cord are a form of uncontrollable activity of the cord and perhaps may be considered epilepsy of the cord.

Epilepsy is teeming with problems, both in diagnosis and treatment. Perhaps the most important is the mode of irritation of nerve cells and the manner in which they work up to the climax which makes them fire off these abnormal impulses. This problem must be biochemical and can only be solved in the laboratory, presumably by a living tissue analysis and other similar methods. At the same time,

new drugs must be developed which will help us to control the currently intractable case, as well as minimising the side effects. From the clinical standpoint it behoves us to examine every epileptic with a view to whittling down that group of cryptogenic cases and developing new tests which will enable us to cut it down still further.

Now what about epilepsy in Rhodesia? It is almost impossible to estimate the number of patients that there are with this condition in the Federation, but there are certainly well over 1,000 amongst the Europeans alone. There is, however, no society which takes an interest in these people, either by helping them to understand their condition or by assisting them with employment. Most of the patients, however, get by quite well in society. They have learned that by continuously taking their sedation they can keep themselves free, or almost free, of attacks and manage to make an adequate living. A number, however, really need help. Sheltered jobs, sheltered education and sheltered homes are required, not to mention education of their friends and relatives to help them understand their condition. There is at present in this country no organisation which does this work, and sooner or later we will need an organisation which can do for epilepsy what RAPT does for tuberculosis.

SUMMARY

(1) Confusion surrounding the use of the word epilepsy with other equivalents is discussed and an attempt is made to show how these various terms used synonymously, in fact, vary immensely in their meaning and often bear no relationship whatever to the neurophysiological and encephalographic findings.

(2) A discussion of seizure patterns in relation to the focus of origin discloses the true meaning of the terms.

(3) Fifty-seven cases of epilepsy are presented and discussed from the aetiological standpoint to conform with the ideas presented above.

(4) The concept of spinal epilepsy is introduced.