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The innocence,

Sleep that but is up the rowelled slave of care
Heckath of each day's life, sore labours bath,
Balm of hurt minds, great nature's second course,
Chief nourisher in life's feast.

MODERN PROBLEMS IN NEUROLOGY

To
THE MEMORY
OF
DR. J. HUGHLINGS JACKSON

MODERN PROBLEMS IN NEUROLOGY

BY

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PREFACE

This volume contains a series of papers dealing with present-day neurological problems which have appeared during the last few years in various journals at home and abroad. Included therein are the Croonian Lectures delivered before the Royal College of Physicians in 1925, the Harveian Lecture given at the Harveian Society of London in 1926, and others delivered at the Annual Congress of the American Neurological Association (1923), at the Joint Meeting of that Association with the Neurological Section of the Royal Society of Medicine (1927), and under the auspices of the British Medical Association (1927). For permission to reprint a number of these I am indebted to the Editors of *Brain*, *The Lancet*, *The British Medical Journal*, *Archives of Neurology and Psychiatry*, and *Transactions of the Ophthalmological Society*. Others have been published in my own journal, *The Journal of Neurology and Psychopathology*.

Though all are recent, each has been submitted to critical revision, and some have been modified to no little extent. New material has been incorporated in most instances. In their present form they embody the results of my study of the problems concerned derived from investigations spread over a number of years. The subjects selected for the book have been chosen because of their importance and of the attention and interest which they continue to evoke; and while no one who has worked at them is likely to claim that in every instance full solution is in sight, in some at least it seems this desirable end is within measurable distance of being attained.

14, HARLEY STREET, LONDON,
1928.



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CHAPTER I

THE EPILEPSIES *

What is Epilepsy? What Constitutes a Fit? Process of Discharge in a Fit. Neural Site of a General Fit. Type of Movement in a General Fit. The Epileptic Aura. The Question of Consciousness in Epileptic Fits. Relation of Epileptic Symptoms to the Cerebral Circulation.

The condition commonly called epilepsy, a "disease" characterized by fits, is familiar to every practitioner; age-old in the history of medicine, it has been claimed as a therapeutic preserve by each successive brand of healer, from the sacerdotal to the psychoanalytic. As has been the case with other common nervous ailments, from time to time there has been a kind of spurt in the race for a panacea, followed only too often by a reaction after the effort; and to-day various rival and mutually contradictory methods for dealing with it are being advocated, the efficacy of no one of which can be confidently and constantly relied on.

To obtain precision of treatment the physician must know the nature and etiology of the morbid condition with which he is dealing and the mechanism of production of its symptoms. Now while the external manifestation of an ordinary epileptic fit is apparent to everyone, its inward or neural genesis and progression offer problems of great difficulty and complexity, on which the outward phenomena shed at the best but an imperfect light; from them we are compelled to infer a mode of physiological activity our theories of which are to some extent speculative, while their transience is such that the opportunity for objective study is often too brief to be adequately seized. Epilepsy is a condition superficially easy of diagnosis but etiologically and pathogenically obscure, and wide excursions in neurophysiology and experimental

* The Annual Harveian Lecture delivered before the Harveian Society of London on April 22, 1926. Reprinted from the *British Medical Journal*, July 3, 1926.

neuropathology have to be taken to gather such relatively meagre data as are at our disposal in this connexion. Even if a psychogenic origin be assumed or proved for one or other of the epilepsies, the physiological problem presented by the clinical symptoms of the fit is left untouched. To say that a given fit is psychogenic does not serve our purpose; in the words of Hughlings Jackson, "all psychological explanations of physical inabilities are merely verbal," and, as anyone can see, a fit in the ordinary sense is a physical thing.

I propose in this chapter to discuss some general and physiological questions connected with epilepsy. Our first is concerned with nomenclature and definition.

What is Epilepsy ?

In the parlance sanctioned by long usage epilepsy is understood to be a "disease" the outstanding symptom of which is the occasional fit. One has only to mention this conception to realize forthwith its thoroughly unsatisfactory nature. There are many organic nervous states of which epileptic fits are a symptom; in fact, the epileptic fit is nothing else than a symptom. No such *disease* as epilepsy exists, or can exist; even by resort to such epithets as "essential" or "idiopathic" we in no sense add a term of differentiating quality, since the pathological element constituting the disease-component (whatever it may be) is not thereby expressed. Further, assuming that a person who suffers from fits is the subject of a "disease" which may as well be called epilepsy as anything else, we are unable to say where it begins and ends. As is known to every clinician, a person may have one fit only in the course of his life, or a few, at the widest intervals, as in the case of a lady who had an epileptic fit at the age of 14, when her period of reproductive activity commenced, and another at 44, when it closed. To term such cases epileptic in the accepted sense is clearly unwarranted, but the nature of the attacks may show that they cannot be called by any other name if the word is to retain descriptive value. Other patients exhibit at the outset attacks justifiably regarded as faints, which subsequently merge by ill-recognized degrees into the epileptic category. And it is scarcely necessary to point out that ordinary major epileptic fits are often for a time the sole discoverable indication of incipient organic cerebral disease—vascular, inflammatory, toxic, or neoplastic.

Nor is the problem of delimitation easier in respect of the clinical

varieties known as Jacksonian or epileptiform, and minor, or petit mal. By objective observation on a single occasion of the extrinsic characters of a series of muscular twitches it is practically impossible in many instances to decide whether they belong to the category of Jacksonian epilepsy, or are rightly described as myoclonus or paramyoclonus, or as tic; additional data not concerned with their objective features are often requisite for an exact diagnosis. Most of us have in recent years become familiar with curious varieties of tonico-clonic involuntary movements as a sequel to epidemic encephalitis which some continental observers are already describing as "striatal epilepsy," as though (vain hope!) the solution of their pathogenesis were thereby facilitated; and many practitioners must know of cases classed by them as petit mal in which they have never been sure whether the clinical appearances really justified that terminology.

In fine, we have been content to classify fits by certain extrinsic characters or qualities mainly because of our relative ignorance of the intrinsic features of the disorder of neural function exteriorizing itself as a fit.

What Constitutes a Fit?

When next we examine the connotation of the word "fit" we find ourselves in no less a quandary, for in the ordinary acceptation of that useful expression it stands for conditions as widely separable as a convulsive movement limited to a segment, or part of a segment, of a limb, and highly elaborate co-ordinated acts of the whole person, sometimes of considerable duration, comprised under the term "hysterical fit." The word cannot even be restricted to phenomena of a hyperkinetic class, for it can legitimately be employed to designate certain attacks of which motionlessness is the prominent feature (see page 42). Put in another way, "fit" and "convulsion" are not synonymous, and the expression "convulsive fit" is not tautological.

Since, then, it is used to comprise attacks varying enormously in type, degree, and duration, not to mention accompaniments, it might be supposed to have become almost meaningless, yet its appropriateness need not be impaired if adequate restrictions are made, and if we can trace the action of some definite physiological process behind its polymorphic phenomena.

General consent, I imagine, will be given to the view that nerve-cells in a state of health store up energy in potentia derived from

fit ← *hinks*
down
control
at cal.

the specific nutritional material they absorb or ingest, and that in functional activity normal movements (the reference is solely to motor nerve-cells in this connexion, although of course the view applies to all varieties of cell) are the outcome of liberation of this energy in kinetic form, to the accompaniment of expenditure of cytoplasmic material. Such normal liberation of energy is orderly, moderate, and controllable. We can, however, readily imagine a condition of motor nerve-cells brought about by abnormal nutrition, and resulting in the development of a state of high tension and instability, whereby they will discharge suddenly, with an excessive liberation of energy, either when a postulated maximum of disequilibrium has been attained, or when a sufficient stimulus reaches them.

Following the profound discussion of this matter by Hughlings Jackson ¹ in his Lumleian Lectures of 1890, I believe that the essence of a fit with convulsive movement consists in the exaggeration of a normal physiological process—that is to say, in sudden, excessive, and yet purely temporary liberation of kinetic energy in a series of motor nerve-cells, the visible consequence of which is a sudden and excessive development of many movements at once. On this hypothesis it is clearly possible for any constellation of nerve-cells in the nervous system to become highly unstable or overcharged, and to discharge accordingly, so that the term “epileptic fit” might be employed, and legitimately so, for any paroxysmal symptoms attributable to the process outlined above. Nevertheless, at this point clinical expediency steps in, to warn us not to extend the meaning of the term unduly, even if only theoretically. Avoiding, therefore, too comprehensive a conception of the word, we are at the same time arbitrarily limiting its designation when we confine “fit” to a hyperfunctioning of *motor* cells. The clinical characters, on the other hand, of suddenness, disorderliness, “caricaturing,” and excessiveness are cardinal features resulting from the physiological process, and without these no symptoms can be justifiably regarded as belonging to the category of “convulsive fit.” This contention will of itself serve to exclude a considerable number of involuntary nervous movements which are the sequel to release of neural function. None the less, as already indicated, to draw clear distinctions between minor types of involuntary movement is often far from easy, especially if the characters just mentioned are not conspicuously present.

The Process of Discharge in a Fit

Our next question concerns the determination of the discharge, and its radiation or spread. If we take an epileptiform or Jacksonian fit, we must concede that the focus of pathologically altered cells may be very small. For example, a patient may suffer from Jacksonian fits which always commence by twitching of the same thumb and forefinger; between the attacks he may be perfectly able to move that thumb and finger voluntarily, hence there can be no material structural defect of the corresponding cortical motor nerve-cells. Some pathological condition, however, in the immediate vicinity leads at intervals to such alteration in their cellular nutrition that they become unstable and prone to discharge "spontaneously" once adequate provocation arises. As already hinted, we can only speculate as to what this stimulus is, but three possibilities suggest themselves: (1) it may be afferent or sensory, setting off the charge as the detonator fires the powder of the cartridge; (2) it may be intracellular, as though there were "spontaneous combustion" when a maximum of tension and instability is reached; (3) it may consist in removal of some inhibitory factor normally restraining the cell from activity. If this third possibility is entertained the problem is merely put a step further back, for we do not know what neutralizes the hypothetical inhibition and leads to decontrol.

Once the "discharging lesion," as Jackson termed it, is in action, the tendency is in the great majority of cases for its effects at once to spread to neighbouring linked physiological groups of nerve-cells, so that in the course of a very brief space of time collateral cells are in discharging activity. It is in no sense likely—indeed, it is hardly possible—that these are anything else than healthy and normal, hence the important conclusion is reached that very little of a Jacksonian fit is directly due to the discharge of abnormally nourished neural units, and that by far the greater part of it is produced by consecutive discharges of normal stable cells. If this be granted, the radiation of an epileptiform fit is entirely a physiological phenomenon, and offers proof of the fact (for which there is abundant evidence otherwise) that healthy neural mechanisms may become epileptogenous, without being in the remotest degree diseased.

In the case of the general, or major, epileptic fit the circumstances cannot be materially different. While on occasion a local

commencement (turning of head and eyes to one side) is indicative of the cell groups first implicated in the liberation of energy, in many instances universalization of the convulsion is almost immediate, and the fit is said to be severe. The supposition here is that the initial discharge is particularly sudden and powerful, and that many series of bilaterally situated cell groups have become unstable and, as it were, explosive. With a great quantity of this explosive material radiation through various neural levels evidently occurs with extreme rapidity, and Jackson's words contain no exaggeration :

"speaking figuratively . . . there is the mad endeavour of the highest centres to develop the maximum of function of every part of the body . . . and of all parts at once ; the phenomena of a very severe epileptic fit show that this endeavour is nearly successful ; the patient is almost killed by the paroxysm, and is nearly dead (deeply comatose) after it."

The Neural Site of a General Fit

It is of interest from the standpoint of neurophysiology to determine if possible the neural site of a general epileptic seizure, a question closely connected with the further matter of the type of movements in a fit. This latter subject is examined later, and is to be considered in association with what is now stated.

If we agree that the property of excessive and uncontrolled discharge is common to all cell groups on occasion, whatever their neural situation, it follows as a corollary that at any of the physiological levels of the nervous system epileptic phenomena may make their appearance. Owing, however, to the intimate physiological integration of these levels the symptoms can scarcely be confined to one only unless the initial " discharging lesion " is mild in degree and incapable of overcoming such resistance as collateral groups offer to excitation. Thus, as already indicated, in the case of a severe fit the chief feature is the rapid spread of the excitations through various levels and to differing mechanisms. It is probably thought generally, or is at least a tacit assumption, that ordinary major fits are the outcome of discharges of cortical nerve-cells throughout the middle physiological level of both hemispheres (the rolandic motor cortex)—as far, that is to say, as visible movements of trunk and limbs are concerned. The view held by Hughlings Jackson was that general epileptic seizures begin in the frontal lobes (his " highest level "), whence the underlying process spreads to and implicates the middle level. Whether this is so or not is

extremely difficult to decide; no constant symptom referable to release, or stimulation, of function of the frontal lobes can be confidently diagnosed; indeed, unless we accept turning of the head and eyes as a frontal symptom (which experimental evidence suggests it may be), no other positive symptom of an ordinary epileptic seizure can be said to have a frontal localization by itself, certainly not the loss of consciousness. It is true that various clinical and pathological data may be taken to indicate that a large part of the brain in front of the precentral gyri subserves motor functions (see page 232); but any motor phenomena its excitation might conceivably produce (with the exception of the not constant head and eyes movement already alluded to) are possibly at once submerged by the spread of the physiological process to the middle level centres. (This point is also referred to below under the heading of "Type of Movement.")

As far as the familiar motor area of the rolandic cortex is concerned, from the physiological standpoint it is electrically excitable, and from the anatomical it contains the Betz cells, which are the origins of the pyramidal or corticospinal tracts running to the ventral horn motor cells of the spinal cord. These corticospinal paths are the chief available routes for cortically initiated movements to exteriorize themselves via spinomuscular units, and in the case of the movements of Jacksonian epilepsy this is the kinetic path followed. When, therefore, the limbs and trunk are convulsed in a severe fit, the idea is that simultaneous powerful discharge of whole groups of motor cells in bilateral rolandic areas is taking place, the result being at first an absolute contention of movement, resulting in relative muscular rigidity—tetanic contractions comparable to that obtained by faradic stimulation of a cortical motor point as long as the stimulus is kept up.

The question arises, however, whether the motor phenomena of a general epileptic fit may not be produced at other levels than the cortical. I have already said that a "discharging lesion," if of sufficient intensity, will scarcely confine itself to one physiological level, but will rapidly implicate others; hence discharges may conceivably be taking place, in this instance, at the spinal level. Epileptic fits can be produced experimentally in decerebrate animals in which the brain-stem has been transected and the cortical motor areas and pyramidal tracts put completely out of action. When only pons, medulla, and cord are experimentally left, fits are still produceable, and convulsions may in these circumstances

develop *sub finem* just as the respiratory centres are failing. Of course, they are possible only because the spinomuscular units are still living, to be convulsed, but the anatomical paths from medulla to ventral horn cells, traversed by the excitations, are not with certainty known. So far as I am aware, strictly spinal epilepsy—that is, in the purely spinal animal—has not been found experimentally, though there is no theoretical impossibility of its occurrence. In view of these and other germane facts the suggestion has sometimes been advanced that the phenomena under discussion are the result not so much of excitation spreading from higher to lower levels as of *removal of inhibition* of higher over lower levels, and that the symptoms of the seizure—that is, the motor symptoms—are produced by release of function; that the epileptic process inhibits controlling centres (say, cortical centres), and that lower centres are “let go.” Hughlings Jackson, however, was so convinced of the far greater complexity of movement representation in the cerebral cortex that he could not accept the possibility of discharges at low levels being comparable to those of high levels; in his own words:

“it would be marvellous if excessive discharges beginning in centres lowest in rank produced fits like those . . . which are produced by excessive discharges beginning in parts of the more evolved centres, the middle motor cerebral centres. . . .”

The problem is one calling for contemplation from every angle, otherwise we are in danger of reaching a premature conclusion. Due attention should be given to the following considerations.

1. While fits of a general kind can be produced in the decerebrate animal, Cobb and Uyematsu² have shown that a much larger dose of a convulsive agent (absinthe, thujone) is required to cause a convulsion in these circumstances than for a similar convulsion in an intact animal, and they conclude that the cortical motor cells are those most easily and readily stimulated by these convulsants. Pike and Elsberg³ also admit the greater susceptibility of the uninjured cortex to experimental absinthe fits, although their investigations have demonstrated the possibility of the occurrence of both tonic and clonic convulsions in animals deprived of both motor cortical areas, provided an interval sufficient for the return of “good locomotor reactions” is allowed to elapse after the initial operation. These fits “did not differ essentially from absinthe convulsions that occur in unoperated animals”; and, according to Cobb and Macdonald,⁴ “the march of events during a convulsion

produced by thujone is very similar to that seen during an epileptic fit." Data of this kind, valuable as they must be acknowledged to be, do little more than substantiate the view already outlined, that in appropriate circumstances healthy neural mechanisms below the level of the cortex may become epileptogenous; they certainly do not validate the conclusion that the neural origin of major convulsions in intact man cannot be cortical. The motor display of major fits is undoubtedly accomplished through spinomuscular units, but more evidence is requisite before the contention can be sustained that the convulsions of organic cerebral disease are, as it were, allowed to occur by removal of some corticospinal inhibition.

2. The evidence associating the phenomena of Jacksonian epilepsy with local cortical disease is sufficiently satisfying to dispel any doubt as to their interrelation; that the march of such fits is in strict accordance with the accepted physiological localization of face, trunk, and limb centres in the cortical motor area, while no representation of an identical kind has been determined in respect of spinal motor centres, is of fundamental significance. Thus I have twice seen Jacksonian fits involving simultaneously only the forefinger and thumb and the corner of the mouth on the same side, a combination readily explained by the juxtaposition of corresponding cortical centres, but meaningless and inexplicable were the epileptogenous site at any lower physiological level.

3. Assuming, for the sake of argument, that the phenomena of epilepsy belong to the category of release phenomena, and that cortical disease only removes corticospinal inhibition, we are faced with the fact that the speculation raises as many difficulties as some may think it appears to dispel. We have to explain the violence and the caricaturing element in the motor discharges, which are unlike those motor phenomena—for example, involuntary flexor spasms—that occur when spinal mechanisms are admittedly liberated from cortical control, as in spastic paraplegia. We have to account for the disorderliness of the epileptic movements, again quite unlike the organized, systematized, scarcely variable movements of released spinal reactions. There is the further difficulty, also, that this postulated temporary release from cortical control is followed by convulsive movements while more permanent release is not.

It seems impossible to avoid the conclusion that there is another element in epileptic discharges than merely a temporary cessation of inhibition from a higher level. We must be careful, indeed, as

to our use of the term "inhibition"; transcortical inhibition is at least as probable as corticospinal inhibition, and the phenomena of epilepsy may conceivably result from decontrol and yet have a cortical site. I have gone more fully into this transcortical question in another chapter, to which the reader is referred (see page 232).

4. Perhaps the most significant point is whether, as a physiological fact, lower (say ponto-bulbo-spinal) centres are capable of exhibiting in action as elaborate motor manifestations as are derivable from the cortical motor ganglia, if the expression be allowed. I have already cited Jackson's view that it would be "marvellous" if this were the case. Unfortunately, this is a matter in regard to which definite observations are unusually meagre and inconclusive. The question is simply this: Are the tonico-clonic convulsions produced by absinthe in the decorticate or decerebrate animal *identical in all respects* with those readily caused by smaller doses of the convulsant in the intact animal? Only minute observation can settle the point. My clinical experience has led me to maintain that in conditions approximating to physiological decerebration in man the fits that may be met with are essentially tonic in character, and not tonico-clonic, as in intact man. The data supporting this conclusion have been recorded elsewhere ⁵ (see also page 24). Such evidence as is available, therefore, in respect of diseased states in the human subject does not warrant the speculation that the march of events in epileptic cases is the same whatever the neural level of the discharges; the sequence of the complicated phenomena of a major attack is not such as can be explained by implication solely of low physiological levels.

The Type of Movement in a General Fit

To ascertain the types of movement seen in a general fit is a matter of observation; their interpretation is matter for discussion.

In the first place it is most important to realize that normal, co-ordinated movements may and do occur along with and separate from those that are called "convulsed." For example, the epileptic patient may, during the fit, make champing movements of the jaws, may smack his lips, spit, make clutching movements at the throat; further, when the convulsive phase is over, and while still unconscious, he may make even more elaborate movements of his limbs, such as plucking at his clothes, etc., all of which movements present the extrinsic features of purposive, voluntary, cortical movements,

yet they are outside his voluntary control. No other conclusion can be entertained than that these have a cortical site; their complexity and purposive, deliberate character exclude any other conception.

Convulsive movements proper cannot be considered as anything else than caricatures of normal movements. They are violent, powerful, disorderly, and generalized, and it is not easy to recognize in them either rhyme or reason. Varying considerably in different individual cases, symmetrical or asymmetrical on the two sides of the body, implicating trunk more than limbs or vice versa, or both equally, they represent a maximum of movement in a minimum of time, and no voluntary mimicry can give other than a faint reproduction of their intensity and severity. The arms may be extended fully and hands clenched, or the former may be flexed; the thumb may be inside the clenched hand, or pressed against the forefinger, or the wrist may be powerfully flexed and the hand more open. The legs may move up and down in violent kicking movements, occasionally in crude alternation, but much more commonly in more or less symmetrical and synchronous flexion and extension. From personal observation of hundreds of epileptic fits, both general and Jacksonian-becoming-generalized, I cannot satisfy myself either that all epileptic fits, roughly speaking, are alike, or that any definite scheme or organization of movement is revealed in their manifestations. On the contrary, I follow Hughlings Jackson in holding that a convulsion is a "contention of complex, and also of simplest, movements. In this contention the individuality of each movement is lost."

It is customary to speak of a tonic and a clonic phase of the convulsions. The former precedes the latter in the average general fit; Jacksonian fits are mainly clonic; a special variety occurs (as already mentioned) in clinical conditions corresponding more or less closely to physiological decerebration, and is known as the "tonic fit," since clonic manifestations are not usually an accompaniment.

The tonic stage of a general fit is one of practically universalized immobility; a number of movements, mutually cancelling each other, are developed with remarkable abruptness and equally remarkable power; the result is a rigid state, a "single big useless movement," as Jackson said, followed by a clonic stage, a series of movements "which do nothing but 'mark time.'" Eventually the patient, exhausted by the severity of this contention of move-

ments, which get slower and slower, sinks into a condition, sometimes not a little alarming, of profound collapse and coma, brief though it may be in reality. On two occasions I have seen a condition in all its essentials as closely resembling death as I am ever likely to observe.

Holding these views of the physiological phenomena of a general convulsion, I can scarcely regard with patience various speculations indulged in by some, to the effect that the convulsed movements are identical with those of the foetus in utero, and that they represent an attempt on the part of the sufferer to retreat again for shelter from an unsympathetic environment into the stillness of the amniotic fluid. I do not consider they have a "meaning" in any recognizable sense; they are of no more "significance" than an explosion of powder. Even in respect of Jacksonian fits, their exaggerated, caricatured movements in no way resemble those called voluntary, while their march presents no point of comparison, but many of contrast, with the co-ordinated movements of a limb in health.

Summarizing at this stage the conclusions reached, largely on clinical grounds, I may state them as follows:

1. The convulsed movements of a general fit resemble those obtained by tetanizing electrical stimulation of the motor cortex (middle level) in their crudeness and severity, and by the action of convulsants on cortically intact experimental animals.

2. In their elaborateness, disorderliness, and violence they do not resemble the involuntary movements known to occur when spinal levels are released by corticospinal disease. The reactions of the spinal animal are orderly, systematized, and scarcely variable with modification of the stimuli evoking them.

3. Combinations of movement are frequently seen in Jacksonian epilepsy that are inexplicable by any known juxtaposition of centres at infracortical levels.

4. Along with the convulsed movements of general fits, "normal" movements may occur; these resemble voluntary movements, except that the element of volition is wanting; their neural site must be cortical, but is not rolandic. It is therefore conceivable that transcortical decontrol is responsible for some of the discharging movements.

5. As far as man is concerned, fits the origin of which is at some infracortical level are not in all respects identical with those of the ordinary major attack.

The Phenomenon of the Epileptic Aura

Consideration of the nature and origin of the epileptic aura is not calculated to lend support to the view that the manifestations of epilepsy are essentially infracortical. While not constant (as far as the mind can subsequently recall the initial stages of an attack), it is when present an integral part of the seizure and may be of localizing value, as every student of the subject is aware. We must keep rigorously in mind the fact that the aura is definable as a sensation, crude or elaborate as the case may be, either referred by the patient to some part of the body or limbs or belonging to one or other of the special senses. Being a sensation, it is a phenomenon of another order than the convulsive movements; it is a psychical thing, in consciousness, and all that can be said of it physiologically is that we believe it arises during functional activity of some central, cortical, sensory mechanism. Because of its being in consciousness its cortical site must be granted (I say nothing here of the speculation that some thalamic activities are accompanied by consciousness of them) (see page 325). In Jackson's clear diction:

"Crude sensations (psychical) and convulsion (physical) are in no way comparable; the comparisons and contrasts are of excessive discharges of sensory elements *during which* crude sensations arise, with such discharges of motor elements *from which* convulsion arises."

I do not know that Jackson, who was always conspicuously careful in his choice of words, was justified in claiming the discharge of sensory elements, constituting in consciousness the aura, to be "excessive," even if "crude." It has always appeared to me that, apart from the occasional normal movements accompanying convulsive movements (alluded to above), the aura is the one thing about the epileptic fit which is not disorderly, distorted, or caricatured. Its crudeness or complexity depends on whether a low-grade or high-grade sensory mechanism is implicated, but we rarely can say of it that it is confused, or made up of conflicting elements, or that it is of exaggerated intensity, or contains any excessive quality.

The problem of its origin is in reality that of hallucinations in general. A sensory aura is a hallucination, of visual, auditory, muscular, gustatory, olfactory, viscerosensory, or any other sense class. Valuable data bearing on cortical localization have been furnished by a consideration of the sensory auras accompanying epileptic seizures occasioned by organic cerebral disease, but we are concerned rather with the general problem of hallucinosis.

The difficulty of deciding whether such phenomena are derived from release or from excitation, is, in my opinion, verbal rather than essential. Completely ignorant as we are of the process whereby function of a sensory mechanism is accompanied in consciousness by what is called a sensation, and, for that matter, of the point in a given sensory arc at which its activation becomes a conscious thing, we are not in a position to assert that a hallucination arises either because a sensory mechanism is "let go" from the control of some hypothetical inhibition, or because the mechanism is being set in action by some usual or unusual stimulus. Even if inhibitory control is at first to be removed (by some equally unknown and speculative process), some excitation might still be necessary to "touch off" the prepared neural unit or system.

That hallucinations are common accompaniments of toxic states scarcely helps us in our search for a physiological explanation of their development. When, as in the case of uncinatc or temporo-sphenoidal epilepsy, a gustatory or olfactory hallucination is often associated with a visual or visuo-auditory hallucinatory state of an elaborate, if certainly brief and fleeting, character, the most we can say is that this state arises in the course of the development of that abnormal physiological process which underlies all the phenomena of the fit, the aura included. Although all auras are psychical, rendering the separation of a so-called "psychical aura" meaningless, there is no good reason for supposing that this sensory component is in some fundamental way different from the motor components, as far as its development in the course of a fit is concerned, and none for imagining that its type, or recurring character, constitutes proof of the psychogenic nature of the epilepsy. Elaborate hallucinatory manifestations occur in persons who are half-drowned, half-hanged, half-suffocated; the hallucinations of uncinatc epilepsy frequently are as definite an index to the existence of organic cerebral disease as the twitching muscles of a Jacksonian case; the *déjà vu* phenomenon and the peculiar psychical condition of reduplicative paramnesia can occur as the aura alike of organic and of so-called idiopathic epilepsy. These and analogous psychical manifestations of epilepsy are analysed in Chapter IV (page 51).

The Question of Consciousness in Epileptic Fits

Some epileptic attacks are attended by loss of consciousness; others are not. Unconsciousness may supervene at the outset,

or develop subsequently ; it may be extremely brief in duration, or may continue for some time after all convulsions have ceased. Further, of itself it forms no gauge of the severity of the fit ; a severe Jacksonian attack may from first to last involve no loss of the senses, while a petit mal seizure may consist of little else than a transient conscious "blank," an "absence épileptique." With this considerable variability in the interrelation of unconsciousness and motor accompaniments the task of correlating their respective causes is not rendered easy.

The evidence for assigning the former to changes in the cerebral circulation is doubtless familiar to most physicians ; it was excellently marshalled by A. E. Russell ⁶ in his Goulstonian Lectures of 1909, which should be read by all who seek an explanation of the difficult problems which epilepsy offers. Starting with this view, a line of argument in respect of the pathogenesis of epileptic symptoms has been developed by the same author, more or less as follows : Alterations in the general circulation afford an adequate explanation of ordinary fainting fits and syncopal attacks ; they also explain the symptoms of more prolonged attacks in which cardiac, vasomotor, and cerebral phenomena can be observed ; the suddenness of some faints, and the conversion of faints into fits, suggest the possibility of the epileptic fit itself having some origin analogous to the disorder of cerebral circulation causing unconsciousness. To this point I shall revert immediately ; for the moment I may say that while transient stoppage of circulation is a plausible explanation of sudden unconsciousness in epilepsy, certain difficulties remain. Consciousness is absent in negative functional cerebral states, such as concussion and coma ; it is also absent during positive, excessive, functional cerebral states—for example, during the period of convulsions. Many clinical instances of the former are unaccompanied by any discharging or convulsive phenomenon ; conversely, a Jacksonian patient may exhibit severe convulsive movements before consciousness is lost. It is undoubtedly difficult to elucidate both negative and positive states by reference to the same process—namely, disturbance of cerebral blood-flow. General cerebral anæmia explains too much, as it were ; with it the excitability of the cortex certainly diminishes, and absence, not excess, of movement should ensue. On the other hand, we might postulate (though we know no special mechanism for it) a *cortical* anæmia, producing unconsciousness, with retention of circulation at lower neural levels, allowing activity of released

subcortical neural systems ; while such a hypothesis might conceivably provide an explanation of the features of the general fit (though, for reasons given above, I do not consider it can) it will not elucidate those of the Jacksonian variety. In view of the occurrence of degrees of unconsciousness in the epilepsies, and of the fact that negative and positive states do not proceed *pari passu* (as far as we can judge clinically), a simple theory of cerebral anæmia is insufficient for the variability of the phenomena.

Relation of Epileptic Symptoms to the Cerebral Circulation

We may, however, in conclusion, pursue this matter a little further. The theory that the paroxysms of epilepsy are a sequel to and determined by contraction of cerebral vessels dates back at least as far as the time of Brown-Séquard. Hughlings Jackson ⁷ himself favoured the possibility of the paroxysm being caused by a local vascular contraction ; he thought that "successions of different movements are developed by contraction of arteries" ; "a convulsive paroxysm is developed by a stronger and more continued contraction" of the same arterial branches as, through coarse disease, are imagined to be producing abnormal and persistent changes of nutrition in the distribution of that artery ; "it is, I speculate, through the arteries that sequence of movements is developed, whether those movements be spasm passing up the arm and down the leg, or whether they be the orderly sequences of movements in health."

More direct evidence, however, has taken the place of speculation. Russell ⁶ emphasizes the importance of the facts that the symptoms of the various stages of the epileptic fit are closely mimicked by those following heart-block in Stokes-Adams disease ; that uræmic convulsions are indistinguishable from those of epilepsy and are almost certainly due to cerebral œdema, consequent increase of intracranial tension being followed by temporary failure of cerebral circulation ; that in some recorded cases temporary cessation of the heart's action (disappearance of radial pulse) has immediately preceded the development of a fit. A number of direct observations of the human cortex during an epileptic convulsion have been made (Horsley,⁸ Kennedy and Hartwell,⁹ Leriche,¹⁰ Horrax¹¹). Sudden blanching of the cortex and pial vessels, and arrest of cerebral pulsation, are at once succeeded by pronounced hyperæmia and venous engorgement during the convulsive movements, according to the mean of the recorded observations. Refer-

ence might also be made to the apparent association of fits with disorder of cerebral circulation in certain cases of Raynaud's disease, of which a number have now been reported (see a paper by H. J. Norman ¹²).

While it would be unwise to ignore the cumulative effect of these differing lines of evidence, I do not think, after having given much consideration to the problem, that they can fairly be taken as showing anything more than that changes in cerebral circulation accompany, in all probability, the march of events in an epileptic discharge. In my opinion Russell is justified in concluding that, even if vascular disorder is causal, it only carries the pathology of epilepsy one stage further. But I am not quite convinced that the association constitutes a *causal* relationship. The cerebral and general circulation is under the control of a vasomotor centre in the medulla oblongata; alterations in the whole cerebral vasomotor apparatus may be caused by dysfunction of that centre. Sudden loss of consciousness may not be of vascular origin, though accompanied by vascular change; the same pathological stimulus may act both on the bulbar vasomotor centre and on cortical neural elements themselves. That this would appear to be the case in respect of unconsciousness following trauma is strongly suggested by recent experiments of Knauer and Enderlen ¹³; and although the analogy with epilepsy is rather remote, it is well not to lose sight of the possibility of the circulatory phenomena of the fit—as observed when the cortex is exposed—being no more than a sequel to a central neural disturbance of function at the level of the medulla.

If, however, we are sufficiently convinced by the evidence sketched in outline above, we should still be at the stage of pure speculation in regard to the question of how transient cerebral anæmia—in some cases only, not in all—causes neural mechanisms to discharge; we should have to imagine a local cerebral anæmia for the phenomena of Jacksonian epilepsy; and I submit that many cases of local and sudden cerebral anæmia, as in monoplegias of embolic or thrombotic origin, are unaccompanied by any discharging phenomena. Nor is the solution of the problem facilitated by ascribing the motor discharges to anoxæmia. Imperfect oxygenation recurring periodically, locally, and seemingly vanishing after an attack—how is such a process to be imagined? And how will any such theory account for the aura of a fit, which, as has been stressed above, unquestionably appears to partake of the nature of a normal, orderly, regularized, non-excessive activation of a given sensory mechanism? Is

neural *activity* accompanied by *anæmia* in the cerebrum of the normal individual?

The truth is that the gulf between the vascular and the neural is almost as difficult to bridge as that between the physical and the psychical. No allusion has been made here to clinical varieties of epilepsy of an unusual kind; if, however, the phenomena of *epilepsia partialis continua* are considered, they may justifiably be used as an argument for the improbability of "discharging lesions" being based on persistence of an abnormal vascular state; they appear to resemble much more those of continuous excitation of a reflex arc by a persisting neural stimulus (see page 23).

It seems to me reasonable to suppose that the crescendo character of the symptoms of the epileptic fit is due to direct action of a neural system on collateral and lower neural systems; if a vascular factor is involved, I have tried to show the possibility of it itself being of neural origin. In some instances this may have a bulbar locus; in others there is as clearly a transcortical commencement for the attack. We come back, therefore, to the view indicated at the outset; any constellation of nerve-cells in the neuraxis may become unstable; hence physiological varieties of fit are to be expected. It is evidently more correct, clinically and physiologically, to speak of "the epilepsies" than of "epilepsy," and this fact by itself should lead us to avoid giving our patients the impression that once a diagnosis of epilepsy is made it is tantamount to pronouncing their doom. On the contrary, increasing latitude in our conception of the condition should lead to modification of therapeutic pessimism.

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CHAPTER II

EPILEPTIC VARIANTS *

Motor Variants: Myoclonic or Regional Epilepsy; *Epilepsia partialis continua*; Tonic Epilepsy; Co-ordinated Epilepsy; Inhibitory or Akinetic Epilepsy. Sensory Variants: Reflex Epilepsy; Sensory Epilepsy; Affective Epilepsy. Psychological Variants; Bad Temper. Visceral Variants: Vasovagal Attacks; Other Visceral Fits.

In the course of practice cases come under notice which present intimate or remote similarities to the phenomena of epilepsy as usually understood, but which do not seem to resemble each other, at least not superficially. For the data in many instances one has to rely on descriptions furnished by either the patient or his friends (there is no reason, of course, to doubt these descriptions are veridical), since development of symptoms during consultation and examination seldom occurs. Study of what may conveniently be termed epileptic variants is particularly fruitful in dispelling from the mind any idea of epilepsy as a self-contained and sharply delimited clinical entity. I pick up a recent book on neurology for practitioners and find the chapter on epilepsy opens with the dictum: "Epilepsy is a dreadful disease." These five words, it so happens, embody two errors, for epilepsy cannot be considered a disease, and experience teaches that the majority of cases are benign. We have laboured for years under the mistaken procedure of elevating symptoms into diseases, giving them names, and forcing new clinical instances into old nosological conceptions. Such has been notoriously the case with epilepsy; so stereotyped is the idea of its being a sinister "disease" that to any given case of "fits" this conception is forthwith transferred—in the absence, that is to say, of accompanying symptoms which may point otherwise. Against this mental habit a protest must be raised; one

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of the best ways of counteracting its harmfulness is by consideration of epileptic variants.

At the outset it is, perhaps, impracticable to dispense with some kind of exposition of what should be conveyed by the term "epilepsy." In a recent Harveian Lecture ¹ (see Chapter I), as well as in a paper ² read before the Assurance Medical Society, certain views commending themselves to me in respect of epilepsy and its connotation have been explained. I may here restate in a somewhat different form what seems to me the appropriate way of facing the problem.

The clinical phenomena under discussion must be considered physiologically; they represent escape from physiological inhibitions. Whether they indicate only release, or also excitation, is immaterial or at least of secondary importance; if an irritative process is to exteriorize itself it must succeed in overcoming inhibition. Escape of function is no preserve of motor mechanisms, as when paraplegic limbs exhibit involuntary flexor spasms, or epileptic limbs are convulsed; or as, indeed, may conceivably be the case with movements thought to be volitional, since even for the manifestation of these release from transcortical inhibition may be an essential preliminary (see page 233). In many epileptic phenomena symptoms of another order are apparent—the aura, if present, is a sensation, therefore a psychical thing, a change in consciousness; sensory systems can undergo release, as in sensory Jacksonian epilepsy (in some varieties sensory manifestations may constitute almost the whole fit). Not a few epileptic symptoms, further, are of a viscerosensory or visceromotor kind. Once the process is initiated, it may, or may not, spread; mechanisms, motor or sensory, or both, on the same level, or on lower or higher, may in turn submit and their resistance be overcome, until in a brief period all are affected, and generalization is then said to take place. Alternatively, the process may be strictly limited, both on motor and on sensory side; the aura, occurring by itself, may constitute the sole symptom and form a larval fit. Again, loss of consciousness, as in numerous instances of petit mal, can develop almost monosymptomatically. Evidently, then, epileptic possibilities are legion; no rigid semiological framework can be fashioned to embrace them all. This becomes still more apparent if we include, as I think we legitimately may, analogous phenomena consequent on excitation or inhibitory release of visceromotor and viscerosensory centres, e.g., those of the medulla. Their clinical

types may seem far removed from epilepsy as ordinarily understood, but they can be considered none the less as belonging to the epileptic category.

Frontal, rolandic, parietal, occipital, temporal epileptic syndromes might be distinguished ; also mesencephalic, pontine, bulbar, cerebellar, and, it may be, spinal varieties. And cannot similar physiological processes conceivably affect visceral neural plexuses and ganglionic collections ? Is "epilepsy" say of the solar plexus an absurdity ? I am far from thinking so.

Common sense compels us, if we entertain this wide conception of the condition, to speak of "the epilepsies," and to disabuse our minds of the idea that one and all must exhibit a somewhat forbidding aspect. Doubtless in many instances we discover at a glance that the epilepsy is symptomatic, as when fits develop during an attack of cerebral hæmorrhage, in pronounced toxicosis, under conditions of intracranial pressure or with perceptible vascular disease ; this fact alone should have taught us to see in epileptic manifestations the action of mechanisms stimulated or released into independent function, and in so-called idiopathic (that is, unknown) epilepsy our search should still be directed towards discovery of the underlying cause, which must exist.

At this point it is perhaps natural to interpose with the objection that the epilepsies would thus be made synonymous with or equivalent to other orders of phenomena with which they do not appear to have much in common. It is not expedient clinically that they should be classed with tics, for example, or with reflex movements and spinal automatisms, or with hallucinations, and so on ; nevertheless, the aura of an epileptic fit is a hallucination, one or other of its movement-complexes can resemble flexor or extensor spasmodic automatisms, and minor, repetitive varieties are often separable from tic only on prolonged observation. What are to be the criteria whereby differentiation is effected ?

In its crudeness, violence, complexity, disorderliness, the major fit can scarcely be confused with any other syndrome, yet parts of it are neither disorderly, violent, nor crude ; the aura shows none of these qualities, and not a few of the movements are co-ordinated, and in no sense convulsive. As for Jacksonian epilepsy, its motor display is largely *sui generis* in respect of both the character of the movements and their march, but slight, localized twitching cannot readily be distinguished from some kinds of myoclonus or of tic. Petit mal is recognized with ease in perhaps

a majority of instances, yet every practitioner knows what uncertainty surrounds the exact diagnosis in others. To establish standards of universal applicability even in regard to the main classes of epilepsy thus becomes almost a Sisyphean task. All of us may think we know a fit when we see it, but we should have great difficulty in endeavouring to specify the criteria which we as clinicians are using. Features such as recurrence, transient nature, interference with the stream of consciousness, are in no way pathognomonic, though they are probably foremost in the mind whenever we think of "attacks" or "fits" of any sort. The phenomena of the epilepsies edge off by easy gradations from the motor convulsive seizure to psychological, sensory, or visceral symptoms of a highly disparate kind, and at the same time refuse to be separated for any physiological reason. Clinical conceptions ought to be based on physiological unities, and if the same process can rationally be supposed to be in action behind these diverse clinical syndromes they belong to the group. No class of symptom in neurology has suffered more from nosological artificialities than that still called epileptic.

I shall take first the clinical types characterized by variations in the motor phenomena, and submit the following classification as possessing clinical usefulness.

Motor Variants

1. Myoclonic or regional epilepsy.
2. *Epilepsia partialis continua*.
3. Tonic epilepsy.
4. Co-ordinated epilepsy.
5. Inhibitory or akinetic epilepsy.

MYOCLONIC OR REGIONAL EPILEPSY

This particular variant is far more common than is usually imagined. It takes the shape of irregular twitches of a limb in multimuscular groups, and in colloquial parlance is often termed "the jumps." "I had the jumps this morning" is a habitual remark. The occurrence of such jerking movements without loss of consciousness is as distinctive as it is little known. Not much complained of spontaneously, careful inquiry elicits a history of their existence often for a longer or shorter time before the first paroxysmal fit. Indeed, in some instances they form the sole epileptic indication, as in the following case.

CASE 1. V. S., female, age 18. Since the age of 14 has suffered practically every morning from more or less violent sudden jerks of the arms, sufficient to throw things out of her hands. Affecting both arms indifferently but not synchronously, these jerks make her knock things off the breakfast table or toss a comb or hairbrush across the room. The legs are unaffected. She has never had a fit in her life, whereas one brother and two sisters suffered from convulsions in infancy, and another brother has had fits since the age of 3. Examination reveals an intact nervous system.

Either unilateral or bilateral, they affect some definite region of the body; sudden adduction or extension of one or both arms, and sudden flexion or extension of the trunk, are the commonest movements. A statistical study by Muskens³ has established their presence at one or other period in no fewer than 103 out of 150 female, and 82 out of 150 male epileptics. A textbook commonplace is to the effect that the familial myoclonus-epilepsy of Unverricht is a great rarity, and the statement is often repeated (cf. Crouzon and Bouttier⁴), yet my experience does not bear out the contention.

The following example is characteristic:

CASE 2. P. P., female, age 15. Has suffered from major fits at long intervals, having had only four in two years. In addition, she shows typical myoclonic jerks of arms and legs in the mornings, throwing things out of her hands or falling suddenly to the floor without a trace of unconsciousness. Examination is negative. A sister also is epileptic, and has myoclonus.

In view of the curious indifference of the average patient to this abnormal condition unless or until major fits develop, its early diagnostic significance ought to be impressed on the practitioner of medicine.

EPILEPSIA PARTIALIS CONTINUA

A second motor variant, doubtless somewhat rare, was described originally by Koshewnikow⁵ as long ago as 1894, and has received but scant attention notwithstanding its physiological interest. It differs from the general myoclonic type in that the twitching is limited to one segment of the body, nearly always a peripheral part such as the wrist and fingers, is practically continuous between the paroxysmal fits, and on the whole partakes of the form less of movements than of irregular, individual, muscular contractions. Among those who have reported cases are Orłowski,⁶ Chorosko⁷ (who suggested the term "*polyclonia epileptoides continua*"),

Spiller,⁸ and Bruns.⁹ I have seen several typical cases, of which one is here briefly related.

CASE 3. E. W., male, age 30. Has suffered from epilepsy since childhood, having on an average several fits every week. For about fifteen years he has complained of irregular twitches of the right hand and fingers, more or less continuous. They are jerky and abrupt, producing usually flexion but sometimes also extension of the fingers, and either flexion or extension of the wrist. Distinct from tremor, they are equally separable from both chorea and athetosis mainly because of their twitching character; in this respect they are classifiable with the myoclonias. The major fits begin in this right hand, becoming generalized with great rapidity.

Of poor mentality, the patient shows on examination no signs of organic nervous disease.

In this instance no objective evidence points to a local lesion of organic or structural type in the left rolandic area; Spiller's patient, however, was a syphilitic. One of the other cases I have seen was under the care of a colleague who considered exploration justifiable in view of the localized phenomena, and suspected a cortical or subcortical tumour. At the operation (at which I was present) no abnormal cortical condition was found. The interest of these peculiar cases consists in the long duration of the twitching, as though due to persistent excitation of a cortical reflex arc, and in the absence of any trace of the exhaustion paresis or paralysis noted with some frequency in Jacksonian epilepsy.

TONIC EPILEPSY

Fits characterized by tonic contractions only, to the exclusion of the clonic element, have long been known. Of their semiology and pathogenesis a description was given in a paper¹⁰ on decerebrate rigidity published some years ago. It was there argued that many so-called cerebellar fits belong to this tonic group, whose physiological localization is in posture-effecting mechanisms of mesencephalic site (mainly). The cortical element in an ordinary fit is supposed to be phasic, that is, clonic, although certain investigations (Pike and Elsberg¹¹) have demonstrated the occurrence of both clonic and tonic convulsions in decorticate (motor cortex) animals, provided an interval allowing for the return of locomotor reactions elapsed after the initial operation. From the clinical standpoint, fits distinguished by the assumption of tonic attitudes and the occurrence of tonic muscular contractions are most likely to develop in consequence of lesions dissociating physiologically

cerebral hemispheres from cerebello-mesencephalopontine levels, and the reader is referred to the article mentioned above for a variety of clinicopathological instances. But brief tonic fits, in some cases not much more than a rather elaborate petit mal seizure, also occur and deserve mention. The following is a recent example.

CASE 4. N. S., female, age 21. When younger, used to have typical myoclonic jerks in the mornings. For the last eighteen months, has had fits about once a week. These attacks, repeatedly observed, invariably exhibit the following features: Ushering them with a cry, the patient throws her head back and falls backward with a crash; trunk and limb musculatures are seized in rigid tonic contraction, arms flexed, legs extended; cyanosis develops while the tonic contraction is maintained for many seconds. No clonic movement has ever been seen. Relaxation then sets in and consciousness returns. A few days ago, her head turning as she fell, she broke the bones of her nose and sustained severe ecchymoses of both orbits.

One of her father's brothers was an epileptic lunatic, and another committed suicide.

Examination furnishes no evidence of organic subtentorial disease.

In all cases of this kind it is perhaps advisable to suspect an organic and localizable basis for the affection, though doubtless such will not always be discoverable.

CO-ORDINATED EPILEPSY

By this term—possibly not the most appropriate that might be devised—the intention is to denote those cases where the movements seen during the attack are co-ordinated and seemingly purposive, exhibiting objectively to a large extent the features of voluntary movements. They may however be aimlessly repeated and in any case do not attain the end to which they may be supposed to be directed.

It is of prime importance to recognize, as is pointed out in Chapter I (page 10), that in many epileptic fits quasi-volitional movements occur along with and separate from those called convulsive. Most of us must be familiar with postepileptic confusional states during which all sorts of automatisms of a complex kind are performed without the patient's conscious participation therein. Similar co-ordinated movement may initiate the attack; I remember a case of left frontal abscess where the patient waved his right arm in circles—as though turning the handle of a barrel-organ—for several seconds ere he fell unconscious and convulsed.

In still other instances the content of the fit is not convulsion but co-ordination of movement, as in the subjoined case.

CASE 5. C. S., male, age 30, has suffered from fits for two years. His attacks occur without warning; he has had one when riding a motor-cycle. In them he turns extremely pale and is unconscious of everything, but never falls; he raises his closed fist and blinks with his eyes, then looks sideways as if to see round a corner. The fit is over in about a minute. He never knows that he has had one, till his wife says, "You've just been having one of your funny do's." A maternal uncle suffered from major epilepsy, also a cousin. The nervous system is organically intact.

As is known, the motor phenomena of petit mal not infrequently are co-ordinated rather than convulsive or myoclonic.

Thus (1) *before, during, or after* the phase of actual convulsion or contention of movements individuality of movement may still be preserved; and (2) occasionally only movements of co-ordination are seen. To me this is a feature of the epileptic seizure of deep significance, for it surely helps to break down any artificial semiological barrier between "functional" and other kinds of fit, and indicates differences of degree only, not of quality, between hysteropilepsy and other epilepsies. Cortical motor centres of the highest level (higher than the rolandic or middle level of Hughlings Jackson) must be implicated if quasi-purposive movements characterize a given fit. Clinically speaking, no distinction can be drawn between the elaborate motor phenomena of a hysterical fit and those of the postepileptic state; both alike represent subconscious or not fully conscious activities released from transcortical inhibition.

These considerations lead to the interesting speculation of the relation of the time factor to the particular form assumed by epileptic disorders of motility. If the motor elements of a convulsive fit could be extended or lengthened in time, would they not possibly show more co-ordination than they do? Is not the *swiftness* of the average major fit responsible for the seeming contention of movement, which results in the "single big useless movement," as Jackson termed it, of the tonic stage? What would happen if it were spaced out a little? The clonic movements, "which do nothing but 'mark time,'" exhibit more than a trace of co-ordination. Were the fit more drawn out, from the standpoint of motion, its features might be so different as to lead to the supposition they were hysterical in nature. The late Sir William Gowers¹² used the apt illustration, in this connexion, of how velocity alters the effect of momentum. "A bullet fired from a rifle makes a

round hole in a pane of glass, which it would smash if thrown against it." Variability in duration of aura and of post-seizure state of automatism is determined by factors of which we have little or no knowledge, and is accountable for phenomena so diverse as to render individual fits hardly comparable one with another, yet the differences are non-essential. After the briefest of petit mal attacks, so rapid as to be scarcely recognizable as such, I have seen a little boy run many yards to hide in any convenient corner—a performance that would have been branded as hysterical had not its real nature been discovered. This *epilepsia procursiva*, so-called, differs in no fundamental way from a fugue that lasts for hours, days or weeks.

INHIBITORY OR AKINETIC EPILEPSY

While the motor syndromes of the epilepsies are ordinarily considered to be essentially kinetic or hyperkinetic, this is in point of fact not universally true. The question arises whether under certain circumstances motionlessness may form a prominent feature of the fit, and must be answered in the affirmative. Faints and syncopal attacks generally, and some kinds of hysterical fit, are distinguished by relative immobility of the patient; in the case of the former the limb muscles are usually somewhat flaccid and devoid of tone; in the latter less so, since rigidity and motionlessness may occur together. I have had occasion to observe certain attacks which can fairly be taken as coming under the epileptic category, in which similar immobility has been remarked. Years ago both Jackson¹³ and Gowers interested themselves in this question; Gowers was of the opinion that discharges in epileptic fits occasionally inhibit, as he found temporary paralysis in some cases after a purely sensory discharge. Jackson referred to cases of epileptiform (Jacksonian) fit in which the patient tells us that his arm "falls dead," there being no spasm in it, whilst the face of the same side is being convulsed. His speculation was that "there may be discharge spreading slowly in a motor centre of the middle level, excessive enough to cause slight after-exhaustion of some of its elements, although one not strong enough to overcome the resistance of lowest motor centres, and thereby to produce actual convulsion."

A peculiar case of unilateral fits which was the subject of prolonged investigation and in which I was able to observe the patient repeatedly during the attacks had as its salient feature the development of complete flaccid palsy of a temporary character after a

sensory Jacksonian aura. It is reported in some detail in Chapter III (page 42). Clearly linked to those mentioned by Gowers, it is classifiable as inhibitory epilepsy.

The topic, however, has in reality wider limits than may at a first glance be apparent. Attacks of one or other kind in the course of which the patient falls or lies motionless, or becomes incapable for the time of volitional innervation, form a perhaps rather heterogeneous group. Consciousness may or may not be interrupted, slight voluntary movements may be possible, slight involuntary muscular contractions may take place. At this point the question of inhibitory epilepsies merges with the problems attaching to trance, catalepsy, narcolepsy and cataplexy—problems, together with a description of various personally observed cases, for a consideration of which the reader may consult Chapter V (page 76). Arguments are there advanced to support the view that some at least of the narcolepsies and epilepsies are interrelated, and that the cataplectic attack, in which the patient sinks to the ground with paralysed and toneless muscles, offers definite resemblances to some of the phenomena commonly called epileptic. This obviates any need to pursue the question further at present, yet I may take the opportunity of alluding to one or two points.

To illustrate concomitance or sequence of narcoleptic and epileptic symptoms the following case may be cited.

CASE 6. A. G., male, age 21. Had some six epileptic fits between the ages of 2 and 7 years, and three or four between 9 and 14. From the description furnished these would appear to have been characteristic major epilepsy. Since 14, has had no more. Recently however, for some months, has developed a different kind of attack, in which he "loses himself," turns white, may or may not fall, sits or lies still, unable to move, without convulsion, tongue-biting or involuntary sphincter relaxation. These turns are brought on by uproarious laughing; if he laughs less immoderately he gets a "pain at his heart," without the attack following. No signs of organic nervous disease are discoverable.

Here is a clear transition from convulsive epilepsy to inhibitory epilepsy, and I consider the case to be of high significance. The late manifestations are classifiable as cataplectic, the early as epileptic. It is obviously impracticable and unscientific to endeavour to divorce the two from the standpoint of pathogenesis, and the case furnishes us with a good example of how misleading mere nomenclature can become. Another recent case may be reported.

CASE 7. G. P., male, age 52. For the last eighteen months has had what he calls "attacks of trance." The aura is a peculiar smell, always unpleasant and usually likened to that of "burning fat." This olfactory hallucination is of variable duration, and is followed by his becoming "weak all over" and "falling into a trance" in which he turns white, is unable to move, and as a rule is unaware of his surroundings. After four or five minutes he "comes to himself," experiencing at the same time a "peculiar feeling" at the root of his nose, which he has difficulty in describing.

In the instance just given we have the same sequence of temporary generalized immobility after a sensory discharge (in this case of the olfactory order), falling in line with those referred to above. It is evident, I think, that the association constitutes no rarity. At the bidding of intrinsic (and as we shall see immediately, extrinsic) stimuli a process of inhibition, not of excitation, is initiated. This is true of numerous cataplexy cases in the narcoleptic category, and also of occasional cases in the epileptic group. Why one physiological process rather than another should be determined cannot easily be explained, and its discussion would take me too far from the subject at present before us.

Sensory Variants

1. Reflex epilepsy.
2. Sensory epilepsy.
3. Affective epilepsy.

REFLEX EPILEPSY

The term is in usage to signalize cases where an epileptic seizure of one or other kind develops on the heels of an extrinsic sensory excitation of one or other order; formerly known as Brown-Séquard's epilepsy (that experimentalist having produced fits in guinea-pigs by excitation of cutaneous "epileptogenous" zones after preliminary injury to the cord), it may or may not be associated with spontaneous fits in a given case. Examples of this type are comparatively scarce, although by now a fair number are on record. In one published by Woodcock,¹⁴ that of a little boy, the remark made by his sister was: "Whenever we undress him he has a fit." Undressed on one occasion in the observer's presence, as his stocking was taken off his right leg he went into an epileptic fit of the classical type, clonic convulsions following tonic, and unconsciousness being complete. I have seen a case in which, when the hat-elastic under a little girl's chin slipped up, hitting the nose,

she went off in an epileptic fit. This accident led to the discovery that flicking or tapping the nose always started the attack, which consisted of dilatation of the pupils, tonic spasm of arms and chest, respiratory arrest, cyanosis, and slight frothing at the mouth, lasting in all less than half a minute. (This case was also seen by Foster Kennedy¹⁵ and is cited by him in a paper alluded to below.)

Another variety of reflex epilepsy was termed by Oppenheim acousticomotor epilepsy, and is distinguished by the patient's falling to the ground on hearing a sudden noise. The following is a personally observed example.

CASE 8. J. R., female, age 19, has suffered since the age of 7 from "falling attacks." She had been knocked down by a taxi two years previously but sustained no actual physical injury. The slightest unexpected noise causes her to collapse to the ground, without unconsciousness, and apparently without any of the usual concomitants of epilepsy, major or minor. So sudden is the attack that she is unable to save herself and has sustained bruises several times. Up to the time of her coming under my observation she has had no epileptic fit. Somatic examination is negative.

A case of this kind is no doubt abortive or incomplete, but it is paralleled and confirmed by one that I have recently been able to see through the kindness of a colleague. In this case, that of a youth, precisely the same abrupt falling attacks have occurred as an immediate sequel to loud and unexpected sounds, and in addition typical major epileptic seizures have developed. I might also cite another case recorded at some length by Foster Kennedy,¹⁵ in which a boy of 17 would fall immediately to the ground on hearing a sudden noise, not by any means always a loud one. He had suffered from convulsions in infancy, and also had exhibited jerking movements of the left limbs and body when he was younger—seemingly a variety of Jacksonian epilepsy. In this case operation over the parietal cortex revealed an old, organized subdural clot, and was followed by gratifying improvement. I have had under observation at the National Hospital a case of *reflex Jacksonian epilepsy*, that of a young man whose fits commence in the right hand and are immediately preceded by a sensory aura of violent tingling in the fingers. This aura and the subsequent fit never arise "spontaneously" but always as a sequence to a sudden extrinsic stimulus of any kind provided it is sufficient to startle him. As a single instance, a horse slipping in the street near him, with clattering feet, sent him off in a fit.

In respect of reflex epilepsy the examples given illustrate the same double process as in the case of the motor variants; an extrinsic stimulus of one or other kind either excites or inhibits. When in consequence of a loud sound a patient falls to the floor it is apparent his posture-maintaining mechanism has undergone inhibition, and the assumption is justifiable that the reaction takes place at mesencephalopontine levels. A certain analogy is permissible with the cataplectic seizures of the narcoleptic (see page 96); under the influence of stimuli of an emotional kind the limbs give way and the patient sinks to the ground, without loss of consciousness.

SENSORY EPILEPSY

That paroxysmal manifestations should be confined to the afferent side is more than a theoretical possibility. We have already seen how much of a fit may be purely sensory—its aura is a sensation, simple or complex, and in some Jacksonian cases not only does a sensation march along physiological lines (precisely as do the motor symptoms) but it also may represent the whole of the disturbance. An example of sensory epilepsy follows.

CASE 9. M. M., age 43, female. Has had for a period of years recurrent attacks, amounting sometimes to as many as twelve to twenty a day; at first occurring mainly at the time of the catamenia, they now bear no relation to it. A sudden feeling of faintness, and a slight flush, is succeeded by a strong olfactory hallucination, always pleasant; "it is just as if I had been passing a perfume shop." The lips then become numb, and this is followed by numbness spreading over the right side of the face, right arm, and right leg. There is a certain feeling of stiffness with the numbness, compelling the patient to make voluntary movements of the lips in order to get this feeling away. No involuntary movement of any kind occurs. The duration of the fit is from three to five minutes. In the course of its development there always appears a faintly conscious background as of voices asking the patient "familiar questions of an ordinary domestic kind."

Careful and repeated examination has not shown that any organic basis for the symptoms exists.

The reader will observe how this case blends with the usual syndrome of uncinata or temporosphenoidal epilepsy (see Chapter IV); as for its actual clinical type, the manifestations are limited, and confined to the sensory system. Some paroxysmal sensory disorders at higher or lower levels (e.g. some of the neuralgias) may well be classifiable with the epilepsies, and the speculation might prove of value as an aid to further observation.

AFFECTIVE EPILEPSY

We may conveniently include among sensory variants numerous cases of epilepsy characterized by the fact of their development consecutive to extrinsic or intrinsic affective stimuli. They are to be distinguished from reflex epilepsy in the more technical sense by the definitely affective element of their excitants, though in another sense one might consider the process a reflex. We should also take note that the variation is not one of epileptic content, but merely of initiation.

"Psychic epilepsy" is a term that has done hard service without ever having been either defined or delimited. "Psychasthenic fits," similarly, have received widely differing connotations (cf. Ernest Jones ¹⁶). It is obvious that expressions of this kind can be applied in psychopathology to fits of a hysterical nature, with which we are not here directly concerned. The reference is rather to ordinary major or minor epileptic fits activated by stimuli of the affective or emotional series. Cases of this sort constitute neither a novelty nor a rarity, and were fully recognized and described long before the War produced them again in some abundance. A typical war case is cited in a paper ² mentioned above; another case under personal observation may be selected.

CASE 10. H. D., male, age 21. Has had typical grand mal attacks for some two years, on an average one every week. A sister, age now 30, has also suffered from typical major attacks for a number of years. One day she fell in a corner of the room in a severe epileptic seizure; overcome with emotion, and with tears in his eyes, he went to her aid, and as he bent down, with words of endearment on his lips, he suddenly lost consciousness and had an equally severe fit on top of her.

This psychogenic or affective epilepsy is as a fact common enough, and instances need not be duplicated. In comparison with what may be termed unmotivated epilepsy, however, their number is insignificant, and I find myself in radical disagreement with Rows and Bond,¹⁷ who assert that in every type of epileptic case a disturbance of consciousness appears, "with which is associated an emotional state," and that "some reaction to express the emotion" can be found in every instance.

Psychical Variants

The term "psychical equivalents of epilepsy" is objectionable for more than one reason; if evidence is forthcoming to suggest

the same physiological processes of excitation or inhibition lie behind the psychical symptoms as behind those of epilepsy in the ordinary sense, then the syndromes are not "equivalent" to epilepsy but are as a fact epileptic. Again, we have seen how much of an ordinary fit may consist of phenomena in consciousness (aura, sensory symptoms), belonging therefore to the psychical series, and it is inadvisable to differentiate psychical equivalents as though they stood for phenomena never found in the usual epilepsies. Indeed, to harmonize "psycholepsy" with epilepsy is not arduous if we adopt the general physiological principles sketched at the outset. Aldren Turner¹⁸ defines "psychical epileptic equivalents" as "the mental phenomena of the pre- and post-convulsive states, when they occur without convulsion or spasm." In my view this is tantamount to an admission of the *identity*, not the equivalence, of the phenomena with other epileptic phenomena.

Thus conceived, the phrase must be taken to incorporate diverse symptoms occurring in petit mal, co-ordinated epilepsy, hystero-epilepsy, postepileptic stuporose and confusional states, and thereby becomes devoid of any specific usefulness. Including dreamy states in the Jacksonian sense, epileptic mania, fugues and ambulatory automatisms, its perpetuation can serve no scientific or clinical purpose.

I take the chance, nevertheless, of referring briefly to an occasional epileptic concomitant which has been supposed to form a psychical epileptic variant.

BAD TEMPER

The occurrence of instances of bad temper in members of epileptic families is noticed from time to time, and has been subjected to statistical investigation by Davenport.¹⁹ In a number collected by that observer the patient was a sufferer from both epilepsy and bad temper, while in other members of the family these occurred either separately or together. The "epileptic temper" has long been recognized and often described (Ribot,²⁰ Binswanger,²¹ Raecke²²), and its exhibitions in prodromal and post-paroxysmal periods are familiar to the neurologist; whether substitution of bad temper for epilepsy constitutes the former an epileptic variant is not to be decided by superficial consideration, since it occurs also in families with insanity as a feature, but without epilepsy. The accompanying diagram gives the details of a family under my observation, in which insanity, epilepsy, and bad temper

are prominent traits. Its interest resides largely in the appearance of bad temper in three members none of whom has ever had an epileptic fit; but the familial occurrence of insanity rather complicates the issue.

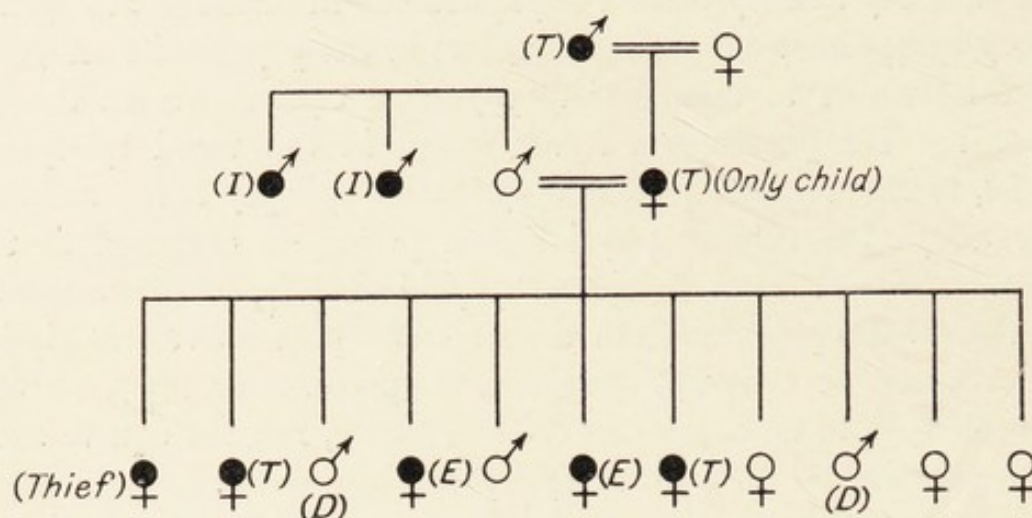


FIG. 1.

I = Insanity.
T = Temper.

E = Epilepsy.
D = Died in infancy.

Davenport's conclusion is to the effect that since violent temper can be shown to occur familiarly without either epilepsy or insanity it should be regarded not as an equivalent of epilepsy but as due to a factor which has its greatest effect when acting on a nervous system that is especially liable to the other.

Visceral Variants

Obscurities arise when, finally, we approach the question of epileptic syndromes of viscerosensory and visceromotor origin. Here we must proceed warily, content to observe if we cannot yet explain. On general grounds, participation of visceral centres in epileptic manifestations must of course be conceded. The sudden pallor of many cases of petit mal, not to mention those of nervous faints, the sphincter relaxation of minor or major fits, are instances in point. As the "discharging lesion" sweeps over different levels it would be curious to find selectivity exercised; the discharge is "brutal," as Jackson said, and does not spare mechanisms belonging to the sympathetic system, in their central connexions. What interests us for the moment, however, is the possibility of epileptic variants the outcome of physiological disorder localized largely if not entirely in sympathetic centres, to the exclusion of sensorimotor phenomena of somatic type. One variety can be

clearly differentiated; others, more fragmentary, can also be exemplified.

VASOVAGAL ATTACKS

This name, which we owe to Gowers¹² (1907), has never received the sanction of common usage; "nerve-storms," "pseudoangina" have a certain vogue, and "anxiety neurosis" a greater one, yet for a number of reasons I prefer that of vagal or vasovagal attack. For years it has been my custom to collect cases falling under this heading, and I²³ have drawn attention to them on several occasions, but the idea is not one, as far as I can gauge, that has taken its place among the conceptions of practice. The expression "anxiety neurosis," as an inclusive term, is misleading, since it is based on the view that the mental and physical manifestations which here concern us are "the inevitable accompaniment of conflict within the personality" (Gordon²⁴). To this assumption I cannot subscribe. That the phenomena do in fact sometimes thus arise I concede, but my contention is that in other circumstances they are adequately described as epileptic variants (in still others, they follow from actual visceral disease). The misconception arises from ignoring the well-established general principle that a given clinical syndrome may be of diverse etiology and of differing localization.

Speaking generally, the symptoms of the vasovagal attack are referable to transient disturbance of function mainly in organs supplied by the pneumogastric nerve, and since implication of the motor vagus is doubtful the probability is they develop from disorder of the dorsal vagal nucleus in the medulla and of the vasomotor centre in juxtaposition. As a result, the patient complains of attacks of palpitation at the heart, difficulty in breathing, a suffocating feeling in the throat, hot flushes, icy coldness, sweating, shivering and trembling, nausea and other epigastric sensations, and often, also, an indescribably distressing sensation of impending trouble, of fear, even of imminent dissolution. One or more of these symptoms, doubtless, may occur as an episodic syndrome in the course of either functional or organic nervous disease, but in the general form already outlined they are periodic and but slightly varying, and their due recognition is a matter of clinical importance. I shall select one or two illustrations out of a large number.

CASE 11. N. J., female, age 28. Has suffered for nine months from attacks which are exemplified in the following excerpts from a description with which she furnished me.

"I was sitting in the garden with another patient at the time and had a quiet feeling as though I did not wish to speak or take notice of anybody or anything, but seemed to be living inside myself. At the same time everything impressed itself upon my mind to an unnaturally forcible extent. I tried to take no notice but to rouse myself and think of something pleasant but could not do so as my thoughts seemed confined; even familiar objects around me, and the birds singing, seemed different and more intense.

"All the time I was conscious of a dread that something was going to happen, accompanied by a fear of death. I had a feeling of slight tingling or electricity all through me and was exhausted, cold and clammy, as though all the blood and strength had gone out of me. Then my heart began to beat rather heavily and quickly, which affected my breathing, making me take my breath in gasps. After that I started shaking all over and then a warm feeling came down over me, like the blood returning, especially in my head and face, and tears came involuntarily. My feet remained cold all the time. When it had passed off I felt better in my mind but weak and shaky in my body, and depressed." By way of amplification it may be stated that nausea and a peculiar feeling in the stomach were also noted towards the outset, and that the flush which developed as the attack subsided was usually accompanied by some perspiration.

A cousin of the patient suffered from major epilepsy. Examination of all the somatic systems has been consistently negative.

CASE 12. C. C., female, age 25. For several months has complained of attacks of "the nerves." On investigation these turn out to consist of palpitation at the heart, difficulty in breathing, sweating, icy-cold extremities, and generalized trembling, together with an anguished feeling of impending death. The attacks vary in duration, from a quarter of an hour to as long as four hours, and after every one there is urgent and almost involuntary defæcation.

Family history and objective examination are alike negative.

CASE 13. F. D., male, age 39, a member of the medical profession. Has suffered from typical major epileptic seizures since his student days. These fits are severe, and are characterized by cyanosis, convulsions, tongue-biting, incontinence, etc. The aura (not constant) consists of a peculiar repetition in the left ear of a word or series of words (not always the same, and not always clearly recollected).

In addition, the patient has frequent "sensations." These also are ushered in by a well-defined repetition of words in the left ear, and consist of a sharp feeling at the heart, "something like a tug," a sense of great difficulty in getting breath, icy-cold extremities, intense fear (possibly of developing a fit); then their place is taken by flushing of neck and face, the heart beats more strongly and quickly, giving a sense of relief, the limbs shake, and pronounced upward eructation of wind always ensues.

There are no signs of organic nervous disease and no structural visceral changes.

CASE 14. I. H., female, age 10. Has had "fits" for the last six weeks. These commence by a sinking feeling in the stomach, associated with giddiness and a form of micropsia; "everything looks quite tiny." Then come severe palpitation, icy coldness and deathly pallor. "I feel as if I were going to die, my heart keeps on throbbing so." Bad headache, cold shivers, and numbness of the right arm and right side of the face round off the attack, which lasts about a quarter of an hour. On examination no organic nervous disease and no visceral cause for the symptoms were discovered.

This patient came again to hospital eight years later (age 18) with a history that the attacks had ceased for a period of several years. In their place another type now occurred, in which the aura was "blindness" and which from its characters was suggestive of migraine.

The above cases are chosen from a large number because they illustrate various points of significance. They may be amplified generally by reference in a little more detail to the symptoms as they unfold themselves seriatim.

(1) The gastric or epigastric sensation is often described as though that organ were moving or turning over, associated with a sick feeling, or as though it were empty or void. "It is a creepy feeling, as if the stomach moves about"; "like being on the waves of the sea"; "as if there were a hollow in the stomach, or a hole"; "as if I were hungry and yet I don't feel hungry."

(2) The cardiac sensation is highly variable, but actual præcordial or cardiac pain is unquestionably rare. Rather the patient notes fluttering, racing, thudding, thumping, of the organ; cardiac arrest is a seeming actuality. In one case, cardiac pain with radiation down the left arm took place.

(3) Respiratory phenomena consist of tachypnoea, less often of bradypnoea, and of unpleasant subjective sensations such as a choking, suffocating, gulping feeling; "my pulse comes up into my throat and chokes me."

(4) Concomitant vasomotor symptoms, equally variable, consist in a coldness like marble—"the coldness of death"—with shivering, trembling, shaking; a cold and clammy perspiration breaks out. Warm or hot feelings suffuse the limbs afterwards. "I feel as if all the blood had gone out of me and that I was left a stone; then I begin to glow all over."

(5) It is of great interest to note that in not a few cases the patient voids large quantities of limpid urine after the attack; in one of the instances cited above defæcation occurred; eructation and flatulence are sometimes accompaniments.

(6) As for phenomena of the psychical series, we note the sensation of *angoisse*, of impending catastrophe, of imminent death. Words of the patients are far more graphic than a laboured description: "Oh, I'm going! I'm going!" "I think it's a death struggle and I say my prayers every time." "I feel, is this it? Is this death at last?" In other instances the feeling is brief and indefinite, yet usually with a sinister content. While fear of one or other degree is common, I have not come across any cases where a definite conscious cause for it existed in the patient's mind.

We also note that there is no loss of consciousness, though frequently a sense of unreality, "as in a dream," closely allied to the "dreamy state" of uncinat epilepsy (see Chapter IV). Sometimes the senses are preternaturally acute, on the other hand; the patient feels abnormally on the alert. Again, particular attention is drawn to the fact that on occasion he is conscious of a sensation of "being unable to move," as if in a trance. "I struggle inwardly, but nothing happens. I try to call out, but I say nothing. I feel if only I could move all would come right." "I hear everything they say to me but I cannot utter a single sound."

This latter circumstance links the condition in some instances to a characteristic of numerous narcoleptic and cataplectic fits—viz., inability to move or to innervate (see Chapter V), and constitutes a phenomenon of inhibition accompanying those of release, as in some other epileptic variants.

Thus with a nucleus of dysfunction in the vagal group concomitant symptoms of an affective kind indicate radiation through other levels, constituting what may not improperly be regarded as an *epilepsy of affective mechanisms*—a periventricular epilepsy, so to speak. It is curiously interesting that the major part of the ganglionic centres concerned with *la vie affective*, and *la vie végétative*, is situated round the ventricular system.

These vasovagal fits frequently arise in cases with a family history of epilepsy, migraine, or insanity. One of my patients suffered from characteristic attacks for a number of years; his mother's brother was insane, and two of his father's brothers epileptic; his mother, and a brother, were both martyrs to migraine. Sometimes no evidence of neuropathic inheritance is forthcoming, as also in many epileptic individuals. I am not concerned here with the appearance of anxiety neurosis as a sequel to "conflict within the personality," hackneyed though such a development is thought to be. Careful investigation fails to reveal any trace of "conflict"

in the cases narrated above ; and the " coitus interruptus " theory is of course inapplicable not merely in the case of juveniles, but also in others in which the practice has never been indulged. To my mind the hypothesis that vasovagal phenomena can in many instances be considered epileptic variants does no violence to the facts and is consonant with what we know of epileptic manifestations otherwise. I do not believe we should expect the reactions of visceral centres in the neuraxis to be identical with those of somatic centres ; allowance must be made for differences, histological and physiological, in the ganglionic collections that are the seat of " discharging lesions." The extended nature of the vasovagal attack cannot of itself form an insuperable obstacle in the way of acceptance of the hypothesis. If a faint is a bulbar inhibitory fit, a vasovagal attack can be a bulbar fit of a less rapid, less " brutal," more co-ordinated kind.

Precaution is desirable, on the other hand, before a given case of this bulbar syndrome can be entered as epileptic. Under very differing etiological circumstances phenomena resembling those that have been sketched can make their appearance. Shock, anaphylaxis, disorders of intracranial circulation, commotio cerebri and so forth, are pathological conditions in the course of which bulbar symptoms can and do arise. Attention was directed by Lévi ²⁵ long ago to the ease with which bulbar disarray (*affolement bulbaire*) develops episodically, while in recent years among the sequelæ of epidemic encephalitis have occurred complex types of respiratory disturbance only a minority of which are classifiable with lowest level (pontobulbar) fits, though few if any are strictly comparable with the vasovagal attack.

OTHER VISCERAL FITS

Other types of recurring seizure in which symptoms referable to central visceral disorder make their appearance have come under notice from time to time, and some are rather difficult to classify, as the subjoined examples prove.

CASE 15. C. S., male, age 11. A very intelligent boy, and a scholarship winner, he has had a series of attacks during the last twelve months which are heralded by deep and incessant yawning, for perhaps half an hour. He turns deadly pale and then starts to vomit, while violent frontal headache develops. The whole thing may last for rather less than an hour.

Objective examinations have repeatedly proved negative. There is no family history of epilepsy, migraine or other analogous condition,

CASE 16. C. L., male, age 25. Has had peculiar fits for about one year, recurring on an average at fortnightly intervals. They are constituted by continual yawning, with constant voiding of urine, and are followed by violent giddiness, headache, and occasional vomiting. There is a family history of epilepsy. No organic signs have been found on repeated examination.

The explanation of these cases is not simple. One might hazard the speculation that they represent some disturbance of third ventricle visceral centres concerned in the regulation of sleep and of water-interchange, with irradiation to visceral centres situated in the floor of the fourth ventricle.

As clinicians, we must be prepared to meet with syndromes, such as these, which do not fit readily into hard-and-fast schemata. In respect of the sympathetic system, as has been well said, neurology is at present groping in a somewhat vague *clair-obscur*. Not enough is known of its normal functions to enable detection of its disorders to be recognized with precision. We may only suspect certain visceral syndromes to be the outcome of functional disarrangement of abdominal plexuses; we still are largely in the dark as regards those derived from changes at higher visceral levels. It is our task, therefore, to observe symptoms, collect data, and collate instances, and imagination may supply the light whereby we shall see eventually how heterogeneous syndromes slip into their place in the physiological puzzle.

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CHAPTER III

INHIBITORY EPILEPSY *

Inhibitory Epileptic Phenomena. Clinical Illustrations. Personally observed Case. Description of Inhibitory Fits. Peculiar Motor Features. Relation to Akinesia in Narcolepsy and Cataplexy.

The fact that many of the phenomena of what is ordinarily understood as epilepsy are inhibitory and not excitomotor in character deserves more attention than it has received. Loss of consciousness—what else is it than arrest of function, indeterminate though its mechanism may still to some extent be? Perhaps the most typical feature of the average case of petit mal is cessation of function in one or other form. The patient stops what he is doing, stares, drops what is in the hand, ceases speaking, falls, sits still and motionless, ignores questions or does not hear them, and so on—whatever the precise formula in a given case the major portion of the syndrome consists patently in the expression of some variety of inhibition. In many severe fits the initial stages illustrate the action of the same physiological mechanism, and so do certain phenomena to be noted occasionally at a later period. For example, loss of sight—darkness, blindness—is not infrequent as an aura; loss of hearing, though less common, occurs similarly now and then; loss of speech, too, may develop at the stage of prelude. Initial muscular relaxation also deserves mention. Not a few post-convulsive symptoms usually attributed to exhaustion are better regarded as exemplifying inhibition.

Of no less interest is the question of stimuli of one or other sensory kind being succeeded not by motor excitation but by motor inhibition. Gowers¹ is one of the few writers who have alluded to this possibility. He was of the opinion that weakness associated with epileptic seizures may in minor attacks be due to inhibition of the same centres as are discharged in severer attacks, and mentioned a case of Jacksonian epilepsy (typical, right-sided) in which

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minor fits consisted of inability to speak or move the right arm or leg, there being no convulsion. More significant for our purpose are two other cases referred to briefly by the same author. The first is that of a woman of 40, probably the subject of organic disease, whose attacks are thus described :

"They begin with 'ticking in the right ear,' and this is followed by a painful sensation 'like hot needles running into the skin,' which passes down the side to the leg and foot and, after reaching the toes, returns up the leg and side and is felt in the arm, hand, and tongue. She mumbles and cannot speak for half an hour. There is no motor spasm, but as soon as the sensation is felt in the leg and arm the side becomes extremely weak, so that, although ordinarily able to walk fairly well, she becomes unable to stand and scarcely able to raise the arm."

The second case, reported with greater brevity, is that of a patient of 48; the attacks consisted in "pins and needles" in the left thumb, which passed up the arm to the shoulder and left scapula, with a sense of contraction in the arm. This became powerless, so that she dropped anything that was in the hand.

These and analogous cases may be taken to illustrate restraint or inhibition of motor centres by epileptic sensory discharges, a phenomenon which, as will be shown later, can be paralleled by other data well known to the physiologist. A remarkable example of the same epileptic syndrome was under my close observation for a number of weeks, and is here narrated in some detail.

Personal Case

The patient was a young unmarried man of 24, by occupation a barman. In all respects both family and previous personal history was negative. He was abstemious in regard to alcohol and had never been the worse for liquor. An intelligent youth, he gave the following account of his symptoms.

Six months ago, without any apparent cause, he began to suffer from curious attacks of tingling in the left hand and arm, and left side of the body, which have continued steadily and are now becoming more frequent. At first these occurred perhaps one or two in a fortnight, but now every two or three days. They may come at any time, but as far as he knows not during the night. In the attacks he never loses consciousness but he feels queer and "regular knocked."

Aura : Of this a very definite description is given, and it has always been the same. It begins by his eyes becoming "misty," not one eye more than the other, or before the other. He can see his way about but he cannot distinguish the forms of objects. He has noticed that

the mistiness is chiefly on looking downwards; on looking to either side he sees "bright flashing stars" in the outer parts of the field. They are not coloured.

This mistiness of vision and these subjective visual phenomena continue for about 15 to 20 minutes before the attack proper begins.

Character : The first symptom is a sensation of tingling in the left fingers and hand. This is variously described by patient—it is as if "he had his hand in a stocking"; as if "it were entangled in string." In the earliest attacks the sensation did not pass the wrist, and he was able to abate it by putting his hand into water, rubbing it, etc. But all the other attacks have been wider in range. The sensation passes up the left arm in a few seconds, then down the left side of trunk rapidly into leg and foot and toes; it then appears (in the more recent attacks) in the left side of the face (lower division). It plays along the lower jaw on the left side, and makes the left half of the tongue tingle. The duration of the sensation varies from only a few seconds to some minutes. When it is over, it leaves arm, leg and face quite numb and "dead."

At the same time as the numbness appears he loses all power in his arm and leg. Anything he has in his hand he will let drop; he will be unable to pick anything up with the fingers. In a second or two the loss of power in the left arm and hand is absolute and he is quite unable to move it. Similarly with the leg, he is completely unable to move it at any joint. The duration of this combined numbness and loss of power may be merely a minute or two, or as long as twenty minutes. When the attack passes off recovery of power is usually simultaneous in arm and leg. He thinks there is little if any loss of power in the left face; it feels as though it were being stretched or dragged up, but it is not so powerless as the limbs.

Sequelæ : Headache (temporal and frontal) usually follows the fit, and may last a whole day. There is no sickness or vomiting.

He thinks that the left side, arm especially, is becoming rather weak, but in any case the degree is slight. Otherwise, he feels perfectly well.

A careful routine examination of the bodily systems showed no abnormality, with the exception that there was possibly a little relative weakness of the left limbs as compared with the right (the patient was right-handed). Sensory system and reflexes were normal. The optic discs were rather high-coloured and the veins full (right more than left), yet they scarcely exceeded physiological limits.

Description of Inhibitory Fits

On three occasions I had the opportunity of observing the fits myself, and a description of their details follows.

I. 12.40 p.m.—When I saw the patient he was lying on his right side, with his legs drawn up, right not so much as left, and with his left arm flexed at the elbow and extended at the wrist. The left foot was distinctly inverted and the toes were pointing downwards. He said that

at that moment the whole of his left side felt numb, including the lower part of his face and round the nose on the left side. He further declared that the sensation had come on when his legs happened to be drawn up, and that he could not move his left leg. When I tested him, he was unable to move the leg at all, and could move the arm only at the shoulder and elbow, and that very slightly.

He exhibited no appearance of distress, no difficulty in speaking, no twitching, and was quite conscious and coherent the whole time.

Testing him quickly, I found his arm rather rigid at the shoulder and elbow, and the wrist and fingers quite flaccid. The resistance to passive movement seemed to be due to defective inhibition of antagonists, a phenomenon well seen in the left leg. The leg was resistant to passive movement, more proximally than distally, and immobile. His face on the left side was distinctly affected, especially in the lower division, for both volitional and emotional movements. His lower maxilla was chattering and he was unable to control this. He remarked then that his tongue on the left side, and the parts underneath his tongue, were numb.

Tested quickly for sensation, there was anæsthesia of the hand and wrist, and of the leg. The limits were undefined. Astereognosis was noted in the left hand, and also very considerable diminution of the sense of passive movement and of position. There was very little if any diminution in appreciation of painful stimuli. All the tendon jerks on the left side were exaggerated, but I failed to elicit an extensor response on the left. The abdominal reflexes were diminished on the left.

12.46.—By this time there was some return of movement in the arm, and also in the leg. He could slowly push his leg down, though he could not lift it off the bed. He was able to flex and extend at wrist and could move the fingers slightly. The position of the foot was maintained. In the attempt to move the leg, defective inhibition of antagonists was particularly well seen. Thus as he slowly extended, the hamstrings were seen and felt to contract spasmodically. The same thing was noticed in the gastrocnemius when he tried to dorsiflex the foot.

Spontaneous left patellar clonus commenced at this point, and when the left knee jerk was elicited prolonged patellar clonus followed. Ankle clonus was likewise obtained, but it was not so well sustained. There was a left flexor response at this stage. The numbness was gradually disappearing from leg and face, but it persisted in the arm and hand. When I was testing his sensation for touch, he remarked that the touch of my finger on his leg and arm and hand gave rise to a feeling of pins and needles at the spot touched.

The facial asymmetry was absent now, and his tongue was protruded straight, although it was tremulous; jaw movement was normal, and the jaw clonus had disappeared.

The loss of sensation to touch remained, and stereognosis was still inaccurate.

12.55.—In the attempt to close his hand, there was some irregular twitching of the forearm muscles, chiefly of the antagonists to the movement. Diminution of muscle sense was not now so marked. By this time the movements of his leg were normal in range but not of normal

strength. He complained that where his limbs had been numb they were now cold, but my hand detected no actual difference in the skin temperature of the two sides.

1.02.—Asked to hold his arms out it was seen that the left arm was oscillating very slightly and irregularly, but he was able to hold it as well as the right.

The reflexes on the left side were now distinctly less brisk than a few minutes ago, although patellar clonus was still obtained. On stimulation of the sole of the left foot very slight flexion of the great toe resulted, and extension of the small ones. The left abdominal reflex was less marked than the right.

There was still slight persisting anæsthesia of the fingers, left, but not elsewhere.

1.08.—All movements on the left were now good in range, though weak generally at all joints. The anæsthesia had disappeared.

II. *Patient again seen in a fit one week later.*—When I saw him, he was lying on his bed with his clothes on. He was perfectly conscious, a little pale, and his lower jaw was chattering incessantly. His left arm and leg were helpless. The arm was flexed at the elbow, and lying across his body. On request, he was unable to lift the hand or arm off his body. Neither limb was stiff or rigid; on the contrary, they were completely flaccid. He complained that all feeling had left the limbs on that side, and that the numbness was extending to the left side of his face. The numbness and coldness began in the fingers and passed up the arm and down the trunk to the leg.

On the left side the jerks were unusually brisk, with definite though not well-sustained ankle clonus. Also there was apparently a left extensor response, though it was difficult to be sure of this; sometimes no movement of the toe followed the stimulus. The left arm jerks were also exaggerated. Both abdominal reflexes were present at this stage.

A minute or so later slight return of power in the limbs was noted, distal more than proximal, and more in arm than leg. At this point sensation was rapidly tested, when no diminution of appreciation of pain on the left side was found, but considerable diminution of tactile acuity. Topognosis also was tested, when definite preaxial localization was obtained.

He then remarked that the numbness had spread to the inside of the left cheek, and under the tongue on the same side. The chattering of the jaw was less, but he said that he had a disagreeable feeling in his throat, as though he were "swallowing a sob."

His sense of position and of passive movement was then tested in the left hand, and it was found that when his third finger was moved he referred it to the first, his fourth to his third, his second to his second. The diminution in sensation continued.

When I was testing his sense of pain again, in the left hand, he said that a prick felt hot as well as sharp. His fingers still felt numb, but the numbness was passing from the rest of him. From this point his left grasp began rapidly to improve, but it was noticed that he could not sustain it owing to defective innervation of the synergic and defective inhibition of the antergic muscles.

A few minutes later the subjective sensations had almost disappeared, except in the fingers. The jerks remained very brisk. The power of the limbs was returning.

III. *Six days later, a third fit was observed in part.*—It had begun with a sensation "like a battery" starting in the left hand and running quickly up the left arm and down the left side into the leg and foot, as well as into the tongue and face (left), and this was succeeded by numbness in the same left-sided distribution. When I saw him, he said he had lost all power over the left limbs. I found that the left side of the face moved less well than the right, and that the tongue was protruded distinctly to the left. The left limbs were limp and useless; voluntary power was practically nil, though there were just the slightest movements of finger flexion.

While I was conducting this brief examination, the clinical condition changed before my eyes (as on previous occasions). The left limbs became rather rigid, the arm moved (involuntarily) into a position of slight flexion at elbow and adduction at shoulder, while the fingers partially closed. At the same time the left leg flexed slightly at hip and knee, extended at ankle, and assumed a position of adduction.

Within a minute or two power commenced to return; movements of the left arm increased in strength and were associated with a sort of coarse oscillation of the left arm at the shoulder—an internal and external rotation of the upper arm two or three times in succession. Passive movement of the left leg evoked a kind of spasmodic alternation of extension and flexion at hip and knee through a very limited range. The deep reflexes were exaggerated on the left side, but both plantars were in flexion, and both abdominals were present and active. Sustained ankle clonus was readily elicited on the left side.

Some ten minutes after the commencement of the attack the patient was rapidly recovering both movement and sensibility, and in a quarter of an hour altogether the above-described symptoms and signs had vanished.

The hyperæmic condition of the optic discs eventually changed into a slight papilloedema, and the question of operation was broached. The patient refused to consider this, and passed from my observation.

Peculiar Motor Features

The case illustrates excellently the occurrence of akinetic or inhibitory phenomena in succession to pronounced sensory discharge, and offers features of considerable importance in respect of cortical physiology.

In the first place, we note that the sensory aura followed strictly the march that is to be expected from cortical excitation, viz. from the fingers up the arm to the shoulder and neck, thence down the trunk to the leg and toes, and at the same time into the tongue and

face. This aura by itself is sufficiently indicative of the cortical site of the physiological disturbance. Secondly, the outfall of motor function was found to vary according to the stage reached in the course of the seizure, the affected limbs being flaccid at the outset, and passing through a period of relative rigidity on the way to recovery. When the arm and leg were completely flaccid they were also powerless, and as tone returned so did a degree of volitional innervation, though the two processes did not advance entirely *pari passu*. It would seem that some degree of return of muscular tonus preceded the other. Of especial physiological interest is the fact, determined in each of the three personally observed fits, that during the process of recovery from the stage of absolute flaccid akinesia the limbs for a time exhibited the phenomenon of *defective inhibition of the antagonists* in a striking fashion. In other words, interference with the normal interaction of synergists and antagonists for the time being existed. This clinical manifestation has long been recognized as liable to occur in cases of hysteria with motor symptoms. Physiologically speaking, hysterical disorders are initiated at cortical or transcortical levels. But defective antagonist inhibition is not confined to cases of that condition. Considerable attention is given in my Croonian lectures to personal observations made in cases of chorea and of athetosis, which have proved that in these states Sherrington's law of reciprocal innervation is often in abeyance and that interruption of the orderly sequence of agonist and antagonist innervation is frequently found (see Chapter X). In the same series of papers evidence is advanced in support of the view that reciprocal innervation can be deranged from the cerebral cortex (see page 234).

If this hypothesis is well grounded, then the appearance of the sign of defective antagonist inhibition in the course of akinetic Jacksonian epilepsy is particularly informative, for it throws light on cortical motor function from the clinical side, and incidentally substantiates the general contention. It cannot be seriously maintained that the motor symptoms of this case have any other origin than the motor region of the cortex.

Actual loss, or diminution, of the deep reflexes was not observed at any point in the course of the attacks in this case. Possibly, had I tested them at the period of maximum flaccidity, it might have been otherwise, but as motor and sensory states were on each occasion first submitted to examination, the reflexes were reached only

after the lapse of several minutes, and this may have been sufficient for a stage of diminution to have passed. However this may be, their exaggeration, the presence of ankle clonus and, on one occasion, of a probable extensor response, clearly point to infracortical release synchronous with cortical inhibition. In this connexion the appearance of an extensor response during a phase of the ordinary major epileptic fit may be recalled. I have not myself been fortunate enough to observe definite loss of the knee-jerk in the course of general fits, but Gowers states it is sometimes abolished immediately at the close of the stage of convulsions.

The development of temporary motor paralysis, without spasmodic discharge, in the course of attacks of the above description, must be assigned to lowered activity of the motor centres involved, that is, to their inhibition, and it is legitimate to assume that the discharge in the corresponding sensory centres has somehow been the cause of this. The likelihood of this being the correct explanation is by no means remote. Under a variety of circumstances powerful stimulation or excitation leads to arrest of function. When a person is "thunderstruck," is "struck all of a heap," in consequence of some violent sensory stimulus, he may be for the moment incapable of movement—he is "paralysed." Reflex action can be interrupted or inhibited by strong painful cutaneous impressions. The only satisfactory way of explaining cessation of the motor phenomena of a Jacksonian fit by ligature of the limb concerned—a procedure known to the ancients—is by assuming an action on sensory centres and inhibition of corresponding motor centres. In the Chapter devoted to epileptic variants I have cited a case in which a strong sensory discharge (olfactory) was succeeded by a trance-like state of motionlessness (see page 29). It is known that a negative phase or refractory state of a given physiological mechanism or reflex arc follows over-excitation. Too loud a sound deafens; too bright a light blinds. We may readily conceive of a refractory or inhibitory phase developing in a motor mechanism if it is suddenly or rapidly assailed with violence by afferent stimulations of an exaggerated character. However antagonistic and mutually exclusive processes of excitation and of inhibition respectively appear to be, they must without doubt be considered at the same time closely interrelated. The most recent evidence pointing in this direction is furnished by the elaborate investigations of Pavlov,² who has conclusively proved how readily inhibition follows stimulation, and how, in respect of cortical function, the

two processes are constantly and continuously interacting and superimposing, one on the other.

This being so, it would be curious indeed were phenomena of inhibition not to characterize one or other of the phases and manifestations of what for want of a better term we call epilepsy. Epilepsy cannot call into being physiological processes that do not exist; at the most it can only distort or caricature the processes of the person concerned. As Hughlings Jackson³ said, "the convulsion is a brutal development of the man's own movements." Analogously, an epileptic development may be a sudden inhibition of the man's own movements.

My chief purpose in discussing the inhibitory side of epileptic semiology has been to bring the latter into line with other recognized clinical states of which akinesia is a, perhaps the, prominent feature. Elsewhere in this volume I have examined at length the clinical symptoms and pathogenesis of narcolepsy and cataplexy, and have stressed the fact that "sleep" is somewhat of a misnomer in view of the observed nature of the clinical syndromes (see Chapter V). These are constituted in numerous instances by generalized motor inhibition, by a state of immobility without loss of consciousness, and in my opinion are often allied to catalepsy on the one hand, and to epilepsy on the other. Instead of rigidly enforcing such criteria as may appear to differentiate all of these, it has been my aim to emphasize their parallels and resemblances.

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CHAPTER IV

THE PSYCHICAL COMPONENTS OF TEMPORAL (UNCINATE) EPILEPSY *

Introduction. Classification. Clinical Illustrations. Description of Uncinate Fit. Hughlings Jackson's Theory. Analysis of Dreamy States. The "Familiarity" and "Unfamiliarity" Types. The *déjà vu* phenomenon. Reduplicative Paramnesia. Application to Epileptic Cases. The "Panoramic Memory" Type. Analogous Phenomena in Normal Persons. Pathogenesis in Epileptic Cases.

A favourite dictum of Hughlings Jackson was to the effect that many different epilepsies are grouped under the one term epilepsy ; if the paroxysms are different, the seats of the discharging lesions, as he called them, must differ. He declared that there are as many different epilepsies as there are different warnings ; put in another way, there are different epilepsies each with its own warning, and some with no warning. The aura is nothing else than the first conscious indication of the commencement of a process that is about to exteriorize itself by signs of the physical series, though it belongs to the psychical series.

Among these differing epilepsies is a type to which the same authority ¹ directed special attention on a number of occasions, viz. that with an aura consisting of what he described as a "dreamy state," coupled, usually, with a crude sensation of smell or of taste, a phenomenon of a different sort from the former. The hallucination of taste or smell may precede, accompany, or succeed the dreamy state ; it may be awanting, or it may occur without the other. As for the latter, it may be extremely brief, almost momentary, or prolonged into the space of a few minutes. Included in the category of dreamy states are such conditions as, "a feeling of having

* Part of the material of this chapter was utilized for a contribution to a discussion on the Psychology of Vision at the Ophthalmological Congress of 1921, and was published in the *Transactions of the Ophthalmological Society*, 1921, vol. xli, p. 116.

been there before," "double consciousness," "feeling of being somewhere else," "memories of things that happened in childhood," "reverie," "reminiscence," "recurrence of an old dream," "a feeling of unreality." Sometimes the feeling is so peculiar and so evanescent that the patient cannot really find words for it; sometimes there are "terrible thoughts" that cannot be expressed.

To those who are unfamiliar with the facts, all this may appear more than a little vague and elusive; nevertheless, if in an ordinarily well-developed form, the phenomena are so striking as to arrest the attention of the observer—subjectively, that is, for their objective expression is of minor importance. When, however, they occur in an imperfectly developed way, it often is a matter of difficulty to recognize them at all, a fortiori to analyse their content. No attempt has hitherto been made to classify these "voluminous mental states," although, as Sir James Crichton-Browne² says, "notwithstanding their diversities they often closely resemble each other for all that they occur in different individuals, and their grouping may correspond to disorder of different cerebral areas."

The reader should understand, to begin with, that in epileptic cases many different psychical states occur as auræ; of these a large number consist of hallucinations of the special senses, constituting a "hallucinatory type." I am less concerned at present, however, with special sense auræ of this kind than with "dreamy state" variants, even if, as is at once apparent, one or other element therein is as a fact hallucinatory. For our point is, that while the psychical conditions to be described may occur singly and separately, it is their frequent association with crude hallucinations of smell and taste that forms the special feature of the group to a consideration of which this Chapter is devoted. The combination, a priori rather unexpected, creates a particular type shown by Hughlings Jackson to be peculiarly prone to develop in cases of organic lesion of the temporal lobe (especially the uncinate region), and often made the subject of study since his first contributions (Anderson,³ Clarke,⁴ Buzzard,⁵ Mills,⁶ Foster Kennedy,⁷ and very many more). Apart from various organic cases that have come under my observation, I have for a number of years specially noted the phenomena as they occur in those belonging to the category of idiopathic epilepsy, i.e. epilepsy of unknown origin; from personal cases of this latter class the majority of the clinical illustrations cited hereafter have been taken.

In the hope of clarifying conceptions and of introducing a little

order into a complex and rather confusing topic I venture to attempt a provisional and empirical classification, and later I shall examine some theories advanced to account for the phenomena.

It seems to me practicable to distinguish four main types of dreamy state, using the term in the Jacksonian sense.

These are :

- I. The "familiarity" or "déjà vu" type.
- II. The "unfamiliarity," "strangeness," or "unreality" type.
- III. The "panoramic memory" type.
- IV. The incomplete or abortive type.

I am well aware that the first three of these may in given cases merge into each other ; in fact, instances of this will be supplied. As remarked above, the combination of one or other of these types with olfactory or gustatory hallucinations constitutes the uncinata epileptic syndrome ; but modifications in the direction of monosymptomatic development of either are naturally to be expected. On the other hand, hallucinatory components of a visual or auditory kind may be added to the syndrome.

Clinical Illustrations

A few specially selected clinical instances will serve better than general description to convey to the reader's mind the salient features of the mental states with which we are dealing.

Type I. THE "DÉJÀ VU" OR "FAMILIARITY" TYPE.

CASE I. W. J., male, age 38, has had several epileptic fits, at long intervals. There are no signs of organic disease of the central nervous system, except as regards the sense of smell, to be referred to immediately. The aura is as follows : "It seems to take me to a set of circumstances in which I have found myself before, or to a place where I have been before. I feel that certain things are happening as they have happened previously, or that people are coming together as at some former time. I never can identify it, however. At the same time, I know where I actually am. Then suddenly comes an overwhelming state as if I were inhaling gas : it draws up my nose and I get a most pungent smell, which is like the smell of gas ; it is always unpleasant. I feel, 'Oh, my God, save me.' It seems the herald of some disaster. About anything seems capable of happening then." Unconsciousness ensues.

Since the onset of fits, five years ago, the patient's sense of smell has slowly deteriorated, though there is no local nasal lesion. He said he could smell neither cigars nor onions. When I tested him, he failed,

in each nostril, with the usual tests, till I gave him asafoetida, when he said, "That's as near as you have got to the pungent smell; I smell it distinctly; it's just like it."

Type II. THE "STRANGENESS," "UNREALITY," OR "UNFAMILIARITY" TYPE.

CASE 2. A. R., male, age 25, has suffered from fits since the age of seven. They average two a month, and consist of typical generalized epileptic seizures. There are no signs of organic nervous disease. The aura is as follows: "A nasty taste suddenly comes into my mouth and everything seems different. It is not a smell; it is a rotten, shocking taste. I can't liken it to anything. It lasts only a few seconds. I often find that I am then making movements with my lips and would bite myself if I did not use my handkerchief. As I look in front of me, things seem different. I can't call to mind what the things are; they are really familiar of course, but now they do not seem so. They seem strange. The whole shop seems strange and unreal. If I can stand still, I am all right; but I feel that I must walk out, and if I can't resist this, and do walk out, I fall unconscious."

A little boy of 11 has given me a brief but lucid and quite spontaneous account of the same thing; its value is enhanced by its concise definiteness and the simplicity of the language.

CASE 3. W. N., male, age 11, has suffered from grand and petit mal for six months. The central nervous system shows no sign of organic disease. Aura: "It is just before the fit is coming; it's a taste like a nasty medicine. When the taste comes, I go like dreaming; all the things go funny like. Once I was speaking to my sister, then suddenly everything in the room went different, as if I was dreaming, and the taste came into my mouth. Then my mouth goes tight, and I don't know any more."

A third instance of this second type is interesting inasmuch as the aura up to the present has constituted the whole attack; there has been no loss of consciousness, and no convulsions.

CASE 4. E. N., female, age 32, has suffered from periodic attacks for the last eight years, averaging several times a month. Her mother's uncle was epileptic; her sister and her brother's son are also epileptic. She describes the attacks as follows: "I am feeling well and happy; all of a sudden, everything around me seems strange and different. I think it funny I should feel like that, because I know I am so well. I've been at the table talking to people, and I've said to myself, I know you, and you, and you, and at the same time they have been unreal and different. They have never noticed anything and yet the feeling has been there. I always feel a curious sensation in my nose; it is a stuffy, burning feeling which goes off very quickly. Then I feel something dreadful is

going to happen ; it does leave me miserable, and I feel I want a good cry." The whole attack lasts not longer than a minute or two. There are no signs of organic nervous disease.

In this case we have an illustration of the "double consciousness" characterizing numerous examples of the dreamy state.

Type III. THE "PANORAMIC MEMORY" TYPE.

CASE 5. T. G., male, age 54, has suffered from fits for seven years. They come at long intervals, sometimes of many months, and are typical generalized epileptic seizures. The central nervous system is normal. The aura is thus described by the patient : "A curious smell suddenly comes into my nostrils ; it is a smell of strong salts, or a strong ethery smell, and it lasts some part of a minute. I then seem to wander back many years of my life to things that happened when I was an absolute child. Trifling things, childish things, come back to me, and I wonder how it is I could remember them. Sometimes I am back many years to some event in my life when I should have done so-and-so, and I didn't. A very short time only elapses in these scenes. It is like a vision. It seems as if I had gone right back and become part of the scene again. I am awake, and yet I seem to be dreaming old things over again." Occasionally the patient "can get out of it by an effort" ; more commonly, the aura issues in the typical epileptic fit.

In the next place, a case combining types I and II may be quoted. The patient is a remarkably intelligent young woman, and the sentences to be given express strictly her own views and are couched in her own phraseology. "Double consciousness" is well exemplified.

CASE 6. E. C., female, age 23, has suffered from fits for three years. There is a history of alcoholism in the family. She has had about a dozen fits in all ; they are typical generalized convulsive attacks. The central nervous system shows no sign of organic disease. For several years before the actual fits came the patient had auræ identical with those which now herald the attacks, without any knowledge of their later significance.

The aura is thus particularized : "I feel I am a different person from my own self, and that I am in a different place from the one in which I actually am. In the attacks I know exactly where I am, nevertheless. The feeling is so confusing. I have a feeling that the thing I am doing, even though it be a trivial thing, I have done before, and a long time before. My idea is that a layer of the brain gives way and another portion of it comes into being."

Again : "At midday I was standing talking to my mother and sister. Suddenly something that my mother said kept echoing in my brain and I felt she was saying something she had said before and everything was exactly the same as it had been before at some previous time. I felt if I stood there the feeling would not go away, so I walked out of the

room and sat on a chair in another room. While the thing was echoing in my head I was afraid; I don't know what of." She was found in a typical fit, a moment later.

Again: "I was writing out cheques in the laundry office, then came a most peculiar feeling of great familiarity with what I was doing; a feeling it had all happened before. I had to go out of the room, to shake it off." However, the fit ensued.

Again: "When I say that in the attack the things round me are different, not real, I mean I see them perfectly well, but they feel different, and feel as I imagine they might feel when being looked at by another person. I feel as if they were giving me the impression which they might give to some one else."

"Directly after the fit, I have an unpleasant taste; I notice it at once. It's not like anything I have ever tasted before. It's a stale taste, not exactly rotten. When I am recovering consciousness the first thing that dawns on me is this taste. It is always the same and it always tells me I have had a fit. If in the office they give me a mouthful of tea, to 'bring me to,' it is spoiled by the taste I feel."

The patient's ideas about the aura in the days when it occurred by itself and had not yet ushered in a fit are very interesting: "I used to feel I had had a previous existence, and that in these sensations I stepped out of myself into the other existence. The things round me did not seem real. I used to feel that I did not belong to the room. I felt as if my brain were leaving my body for the moment. I used rather to welcome it, as a not unpleasant feeling."

Hitherto I have cited cases exemplifying the association of different forms of the dreamy state with crude hallucinations of smell and taste. This latter element, however, may be wanting on occasion, as in the next two instances, though the dreamy state otherwise is quite typical.

"Familiarity" type without taste or smell.

CASE 7. E. B., female, age 23, has suffered from fits since the age of 15. Aura: "I feel as if I were in a dream, or half asleep, though I know exactly where I am and what is round me. I always feel as if I had been having a conversation with some one and they with me; the voices are familiar but I never can quite remember whose they are or what they have been saying. I once went to Kew Gardens; I had never been there before. One of the attacks was coming on just as I went in. I felt at once that I had been there before; it was quite familiar. It made me feel quite miserable, and tears came into my eyes."

"Memory" type without taste or smell.

CASE 8. C. W., female, age 35. Her mother is insane. Her father's father and her mother's father both died in asylums. Her husband is

at present in an asylum ; his mother died insane. Her husband is her mother's sister's son. The patient had convulsions in infancy and fits commenced again when she became pregnant eleven years ago. They average two a month, and are typical generalized epileptic seizures. There are no signs of organic nervous disease. Her aura is thus described : " I go into a curious state in which I suddenly remember all sorts of things that happened when I was a child. They are stupid, silly, old-fashioned things of my childhood. They are things that I have done in years gone by, in other places. I remember silly things like swinging, playing with toys with other girls, and then I go off." There is no subjective sensation of smell or taste.

Type IV. THE INCOMPLETE OR ABORTIVE TYPE.

Mention has already been made of a group of cases where the dreamy state is not fully developed, or is incompletely remembered, or, indeed, where the patient is unable to give definite or adequate information. Larval types of this kind are as a rule, though not invariably, unaccompanied by gustatory or olfactory sensations, a point which is not to be wondered at since the whole syndrome is exceptionally faint in degree even if none the less a prelude to a major epileptic seizure.

CASE 9. M. K., female, age 29, has suffered from fits for eight years. Her aura is an " unexplainable something " : " I call it a vision, but I never can get at it. It is a funny sensation which lasts only a minute ; it is always the same, and if only I could get at the root of it, I should tell you."

CASE 10. W. F., male, age 22, has suffered from fits for two years. His aura is thus described : " I always feel that I have been somewhere, but cannot tell where it is. I keep puzzling over it. Somehow there is with it a feeling of dread, as if some one were going to strike me. I never can find out where it is I have been ; that is the funny part of it." This aura is invariable.

[Description of Uncinate Fit

To conclude these clinical illustrations, and to give the reader a general idea of the evolution and course of an uncinat fit, complete with dreamy state and hallucination of olfactory-gustatory kind, I shall give succinctly the record of one observed personally from start to conclusion. The case was of organic character, the lesion being a right temporosphenoidal abscess.

CASE 11. H. B., male, age 29. Patient was sitting on his bed when I reached him. He was quite conscious, though he looked very distressed. He said : " Hallo, Doctor ; I want you. I have been trying to keep

it back, but I can't. I was playing at draughts when it began. I smell and taste it everywhere; it's stronger than ever."

(He told me the next day that he was sitting in the dayroom, when it began with a feeling of seasickness. He remarked to his neighbour that he felt sick, and began to walk back into the ward, feeling rather giddy. Just as he entered the ward, he noticed an overpowering sensation of bad gas, like the fumes of spirits of salts, which he seemed both to taste and to smell. He thought he was choking; the ward seemed full of it. When he reached his bed, he was slightly sick, and immediately thereafter he heard bells, which were high-pitched, and seemed to be far above his head. He thinks he heard them first in his right ear. They lasted only a few seconds.)

He was then sick, vomiting slightly into a basin which nurse had put between his feet. The expression of his face became more pained, and he caught at his knees with his hands. Then suddenly he smiled faintly, looked up, and said, "Is that you, Porter?" I asked him if he saw anyone, but he made no reply. He then turned his head very suddenly to the left, and said abruptly, "Who's that? Is that you?" He was then sick again, and a moment later the right arm and hand, followed almost immediately by the right leg, began to tremble. A moment later the left arm and hand, to a less extent the leg, were trembling too. This continued for a few seconds, then it ceased.

(He informed me subsequently that the left leg and foot felt very cold, while the right by comparison was hot. He also stated that he thought he saw the friend whose name is given and whom he had known for years, and that he was engaged in conversation with him, as he had often actually been. The whole thing was familiar and real.)

All this time he had been sitting on the edge of his bed. At this point his head, eyes, and upper part of his trunk turned slowly and steadily round to the left in a powerful tonic spasm, till the upper trunk was rotated through more than a right angle. At the same time he gave a long inspiratory cry, and the left face drew up in a tonic spasm. He was put by me on his back on the bed. A characteristic major epileptic attack followed.

As soon as the fit was over, he became restless, began to respire very deeply, a blowing sort of respiration, and at the same time made constant spitting, champing movements of his jaws and lips. He did not actually spit. He kept pursing and pouting his lips, and moving his head from side to side.

In the course of about three minutes these movements died away, and he sank into a drowsy condition.

The heterogeneousness of these selected cases is more apparent than real. In all of them the dreamy state bears a definite relation to typical generalized epileptic seizures; it occurs as an *aura*, and in any particular case is constant for that case. Complex though it be, or subtle and elusive, it nevertheless stands in the same position for the subsequent fit as does an epigastric *aura*, or an

aura of any other simple kind; and we analyse it and assess its localizing value just as we would an aura of tingling in one hand or foot.

Hughlings Jackson's Theory

This is not precisely the same attitude as that adopted by Hughlings Jackson, whose general view of these dreamy states was somewhat as follows. He⁸⁹ believed that all elaborate although morbid mental states arise during activity of centres which are healthy, except for loss of control. The epileptic discharge places the highest or controlling centres hors de combat. Dreamy states are not comparable to ordinary warnings; they are not the result of epileptic discharges of the highest centres, but are owing to over-activity of the next lower centres, as yet untouched by the discharge. I may cite his own words: "In no case do I believe it possible that elaborate states can occur from an epileptic discharge. I believe all elaborate positive states occur from, or arise during, an increased energizing of centres permitted by removal of control of higher centres." He considered a dreamy state not comparable to an aura in the sense that a hallucination of smell is an aura. "It would be a remarkably well-directed and well-distributed epileptic discharge which would give rise to the exceedingly compound mental state of being somewhere else. Besides . . . it is scarcely likely that one thing, an epileptic discharge, should be the physical condition for a sudden stench in the nose—a crude sensation—and also the physical condition for an infinitely more elaborate mental state."

From this summary of Jackson's views the reader will gather that he drew a distinction between *discharging* phenomena and *release* phenomena, placing the aura in the former category and the dreamy state in the latter. As I take it, however, this is an example of a distinction without a radical difference. I include both groups in the epileptic category definitely, and consider that a given phenomenon is none the less part of the total epileptic process because it arises from the action of healthy centres "as yet untouched by the epileptic discharge." For a discussion of this aspect of the problem the Chapter on The Epilepsies may be consulted (page 5). It matters little from the viewpoint of phenomenology whether a given symptom, physical or psychical, is the outcome of excitation or of release. As a further instance of the particular use by Jackson of the term "epileptic" I may

quote his opinion of the occurrence of co-ordinated movements in fits (see page 10): "I would suggest that such movements as those of mastication, of spitting out, and smacking the lips, imply an excitation of some centres for taste analogous to excitation of other centres producing 'balls of fire,' etc. . . . *not an epileptic discharge, which would produce spasm of the muscles*" (italics mine). Here again, according to my views, is more of a distinction than a difference. I cannot agree that a motor symptom, to be epileptic, must essentially be one of convulsion (see Chapter I).

Analysis of the Dreamy States

The odd association of the dreamy state—be it a sense of familiarity or of strangeness, or a string of old memories, or something less tangible—with a crude hallucination of taste or smell is the most remarkable feature of the cases under discussion, more remarkable than the occurrence of either separately. Their synchronous evolution is no mere hazard of cerebral disturbance, but a combination of definite diagnostic significance. The student of the subject is of course well aware how difficult it is to draw a hard-and-fast line between larval states of this "intellectual aura" and warnings so vague as to be practically valueless for our purpose; moreover, he knows that hallucinatory phenomena involving other senses than smell and taste are met with, so that, in spite of my empirical classification, he may still feel the conception is such a medley of psychical fragments as to be unworthy of more serious attention. The definiteness of typical cases, however, and their unvaryingness for any given instance, as a rule, cannot be gainsaid. It is unwise to minimize their import because of inability to deal satisfactorily with minor cases. I frankly admit it may be impossible to say whether a given aura amounts to a dreamy state or not; in suggesting a classification I have offered no definition; but the development of elaborate mental states in association with subjective sensations of taste or smell is an observed fact in both organic and idiopathic epilepsy, and I submit it is one calculated to throw light on the function of certain cerebral mechanisms both in health and disease.

The "Familiarity" and "Unfamiliarity" Types

We may consider these two together, since one is as it were the obverse of the other.

The phenomenon of "déjà vu" (paramnesia, "fausse recon-

Old John and the children, and the laughter and the memories—memories of other childhoods and other sunrises, of other birds that are the same for us, and the same bells that are so different for us ; and then the mother-love, and mother-comfort, and home for the happy feverish limbs, and the long silent dusk on the empty green—

Derby and Dow. Mr. Honey sets out
evidence that exists as to the output
factory with all the logical skill
persuasion of a modern detective
and he ends with a pretty conjecture
own concerning a tea-caddy at Har
has long been a stumbling-block to

naissance"), or better, perhaps, "déjà perçu" (Ballet¹⁰), is undoubtedly of common occurrence in normal persons, as we all know. By it is meant simply the feeling (it is not so much a feeling as a judgment) of the particular set of circumstances or environment in which we find ourselves at the moment having occurred before or been experienced before, on a long previous occasion. In its fully developed form we should be able to state definitely that we have never as a fact been in that particular environment or experienced that particular set of circumstances before. Further, we never actually remember the scene or set of circumstances with which we imagine the present scene or occasion to compare, thereby introducing the familiarity-element; or if we do, it is certainly not at the moment when we first experience the familiarity. If we did, we should be able to grasp the differences between past and present, and the feeling would vanish. We may note, also, as a corollary, that if the feeling leads us to say in astonishment, "This is the second time I have been here," it is never supposed to be the third time.

This mental state is capable of a number of widely differing explanations.

I. *The reminiscence may be genuine*; that is, it may depend on an actual previous experience.

(1) This may be one of *waking life*. Actual past events, dimly remembered, their details forgotten or only partially recovered, may induce the belief that the new state is a repetition; it is an error, but only in part. In my opinion, such an explanation is sufficient for a large number of simple cases.

A friend of mine remarked spontaneously that she frequently experiences the sensation of *déjà vu* when wandering through a ruined castle where she has never been before. Now, in the features that strike the ordinary observer, one ruined castle, with its broken staircases, its silent courtyard, its empty echoes, its dungeons, is most certainly just like another, so that in this instance the reminiscence has its basis in ill-recollected actual previous states. Similarly, to enter a strange drawing-room and be overwhelmed by a sense of familiarity with not merely objects, but persons and conversation, is an experience capable of this simple explanation.

But the previous events on which the pseudo-memory is based, though real, may be unknown to the subject. Abercrombie¹¹ narrates the case of a lady of 80 who, entering what she believed to be a strange house, looked anxiously round one of its apart-

ments and said, "I have been here before; the prospect from this window is quite familiar to me." It turned out that as a little child she had been taken to that very house from a considerable distance, to see her mother who lay dying in it, in that particular room, and that she had never been there since, nor had she any trace of recollection of the visit nor was the name of the village ever known to her.

(2) The actual previous experiences may be those of the *waking imagination*. Illusions of memory may arise from the products of mental activity we call day-dreams, fantasies, reveries. Had Wordsworth's mental image of the as yet unvisited Yarrow resembled the reality more, when at length he reached the river's banks he might have experienced the *déjà vu* phenomenon in all its intensity. Sully¹² says that "a person's name, a striking saying, even an event itself, when we first come across it or experience it, may for a moment seem familiar to us and recall some past like impression, if it only happens to resemble something in the works of a favourite novelist."

I have come across an apposite illustration in the book by E. F. Knight¹³ entitled *Where Three Empires Meet*. Wandering through the land of Ladak, on the borders of Tibet, he was for some time greatly puzzled.

"It all, in a way, seemed so familiar to me. Surely I had somewhere, long ago, lived amid this curious people and in such a weird land as this—but when and where? . . . I felt quite relieved, at last, when the explanation of this mysterious feeling flashed across me. . . . I remembered that when a small boy I had read *Gulliver's Travels*, and that the voyage to the flying island of Laputa had made a great impression on my imagination. I had conjured up that kingdom to my mind just such a perspectiveless, artificial, unreal-looking land as this; and just such a people as these queer Ladakis had those no more queer people, the Laputans, appeared to my fancy."

(3) The reminiscence may have its source in *dream-consciousness*. The latent traces dreams leave in our minds may be stirred by some accidental resemblance, and it is clearly possible they are to some extent answerable for the sense of familiarity experienced in visiting a new locality. Under certain conditions the difficulty of differentiating what has been experienced from what has merely been dreamed is considerable. Cases have been recorded where the chief symptom consisted in the patient's inability to distinguish between the facts of his dream life and of his waking life.

Ferenczi¹⁴ attaches importance to the unconscious dreamings of the mind at night; of these he thinks we are unconsciously reminded in an actual situation. Freud¹⁵ has alluded to the matter in his most popular book, and selects the "unconscious fantasy" explanation, to the exclusion of all others. "Those psychic processes which, according to my observation, are *alone* responsible for the explanation of *déjà vu*—namely, the unconscious fantasies—are generally neglected by the psychologists even to-day. I believe that it is wrong to designate the feeling of having experienced something before as an illusion. On the contrary, in such moments something is really touched that we have already experienced, only we cannot consciously recall the latter because *it never was conscious*. In short, the feeling of *déjà vu* corresponds to the memory of an unconscious fantasy" (*italics mine*). Characteristically, in the example selected Freud works the unconscious fantasy round to a repressed wish.

F. W. H. Myers¹⁶ agrees in recognizing the significance, for the familiarity feeling, of a suddenly evoked reminiscence of a past dream, but for him the important question is, whether the connexion is more than casual; whether the dreamer may have in some *super-normal* way visited the scene, or anticipated the experience. Both he and Sully linger over the possibility, and scarcely conceal their fascination by it, of the hereditary transmission of mental phenomena, instantaneous emotions of the mother imprinting themselves as memories on the foetal organism. "If it were found that a child that was descended from a line of seafaring ancestors, and that had never itself seen or heard of the dark-gleaming sea, manifested a feeling of recognition when first beholding it, we might be pretty sure that such a thing as recollection of prenatal events does take place." Freud's *Traumdeutung* contains several instances of dreams which he chooses to interpret as based on unconscious antenatal impressions. It may be so; but if we explain shadowy recollections by shadowy speculations, we do not advance very far.

II. *The reminiscence may be illusory*, no previous dream or waking experience underlying it; it may be caused by abnormality in perception or recognition processes.

Myers' contention that such explanations are purely hypothetical is curious in view of his own special theory that the cause (or rather one of the causes) may lie in a double perception of the present moment by the subliminal and the supraliminal self. Freud's dismissal of such explanations, without discussion, is the more

regrettable because his own view fails obviously to explain such *chronic* cases of the condition as have been described by Pick,¹⁷ Coriat,¹⁹ and others under the heading of "reduplicative paramnesia." Pick's patient was an educated man, who had suffered for many years in the following way: if he was at a social gathering, visited any new place, met a stranger, the incident (with all its attendant circumstances) appeared so familiar that he was convinced of having received the same impressions before, of having been surrounded by the same persons or the same objects, under the same sky and the same state of the weather. If he undertook any new occupation he seemed to have gone through with it at some previous time and under the same conditions. In Forel's¹⁸ case, "every position, every attitude, every word that anyone spoke was to his mind a repetition down to the minutest details of an original which he localized in the past." Coriat mentions cases of alcoholism, general paralysis, and senile dementia, in which the phenomenon was equally chronic. Of several examples I have seen, one was in a case of general paralysis, and another in that of a chronic epileptic, who furnished me with interesting details.

Reduplicative Paramnesia in Epilepsy.

CASE 12. A. B., male, age 39, has suffered from major and minor epilepsy for eleven years. No evidence of organic nervous disease can be found. In my consulting-room he described his condition as follows:

"I seem to be constantly in a state which is a panorama of my past life. At this moment, I feel that I have said all this to you before, exactly the same. I do my best to shake it off, it is so uncomfortable. Everything that you are saying to me I feel you have said before. It is exactly the same with my wife's sayings and doings—in fact, with everything. My memory seems too good. I write down all my engagements now because I feel I have really carried them out recently. When I am talking business it is just the same. Yesterday I rang up a friend and asked her to come to lunch; no sooner had I done this than I felt strongly I had already had her to lunch, a few days ago, so I immediately rang her up again and apologized for my error."

In this case the phenomenon had been present for many months and applied universally to the experiences of every passing minute. To assign these symptoms of my own and other recorded examples in each particular and ever fresh instance to recollection of unconscious fantasies is in my opinion frankly impossible; the hypothesis breaks down under their weight.

III. Various *psychological theories* of perceptive or recognitive dysfunction may now be alluded to.

Lewes ²⁰ believes that a thrill of emotion diffuses itself over the field of consciousness and obliterates the landmarks whereby new and old would be distinguished.

"We have only to attend to our sensations to be aware that they do not cease with the cessation of the stimulus—the tones keep sounding in our ears. . . . Now when a wave of feeling has swept through us, and another similar though fainter wave succeeds, the secondary feeling will naturally be taken for a vague remembrance, the resemblance between the two being accompanied by a difference in intensity which throws the second, as it were, into the distance."

Lalande ²¹ assumes the possibility of a singular and almost indefinite acceleration of thought. It is possible for the human mind to represent to itself in a few seconds a series of conscious states whose ordinary subjective duration would occupy several hours, or, indeed, much longer. Hence a moment of distraction, in between two perceptions of the same place—a moment during which the individual's thoughts summarize a very much larger subjective period—will explain the occurrence of the phenomenon.

Ribot's ²² explanation is in a way not dissimilar. He emphasizes the fact that in health, when recollection occurs, not stimulated by the actual presence of the object, the originally received impression is reproduced in the form of an "image." When the phenomenon of *déjà vu* occurs, the image of the state, formed a moment after the real state, somehow gives to the latter the character of a repetition. How is this effected? Ribot thinks that the image formed is so intense that it imposes itself as a reality on the mind. "The real impression is relegated to a secondary place as a recollection; the image becomes the reality and the reality the recollection. . . . The illusory state does not efface the real impression, but as it is detached from it and produced by it, it appears as a second experience; it appears to us more recent than the other, as indeed it is." Ribot, however, does not pretend that his theory is the only one possible.

Another theory for which there is rather more to be said takes cognizance of the occasional occurrence of a separate "familiarity-quality" or "familiarity-feeling" (*Bekanntheitsqualität, Bekanntheitsgefühl*) as a psychical phenomenon attending perception. This "familiarity-quality" transforms perception into recognition erroneously. It does not depend on any memory process, or on what one might call probable inferences, or on the reproduction of similarities. In the pathological cases of reduplicative param-

nesia already alluded to the several parts of one and another situation are as a fact entirely strange to the patient concerned and yet every one of them is felt by him to be completely familiar. To explain these admitted data Störring²³ suggests that the phenomenon under immediate consideration is due to reproduction of an idea of a "previous state of the self." Only in this way, as he believes, are we able to understand why the total present state may be apprehended as familiar; it follows naturally from the content of the re-presentation of a previous state. If this re-presentation results in recognition, then the assumption would be natural that the recognition is based on association of a previous state of the self with previous perception of the object.

Application to Epileptic Cases

Among these various kinds of explanation of the occurrence of paramnesia in presumably normal persons some appear more applicable than others to diseased conditions. From so much psychological uncertainty and conflict the clinician cannot be expected to derive much assistance. It is conceivable that the manner of the arising of *déjà vu* may be different in normal and abnormal circumstances respectively, though its qualities are identical in the two.

In the cases we have been studying we have seen that the phenomenon is of not infrequent occurrence as an aura to certain epileptic fits, and that whatever be the environment of the patient at the moment of onset, it is this environment, or a part of it, that becomes impregnated with "familiarity-quality" (cf. in particular Case 6 above). Among other cases, as we have also seen, is a group in which actually familiar environments become at once unfamiliar, strange, different, unreal. In others, too, we have found that ere the patient loses consciousness he finds himself in a curious "double" mental state; he knows where he is and at the same time feels as if he were somewhere else; familiar objects round him become unfamiliar, while the "somewhere else" becomes familiar, and he feels he has been there before.

Now if we accept Störring's suggestion that the basis of the familiarity-feeling is a previous state of the self, we are not faced with the difficulty of explaining how similarity is to account for it when the circumstances that form the setting of each fit are bound to vary enormously. Arguing by analogy, we might say that the strangeness, the unfamiliarity, the unreality, of which so many of

these special epileptic patients are conscious, is the expression of a defective state of the self, independent of the absence of similarity-reproduction as far as the environment is concerned. This much is clear, I consider, that the facts of clinically observed cases are the outcome of intrinsic and not extrinsic processes ; as a consequence of abnormal cerebral states, momentary or more prolonged, mis-perception or mis-recognition develops, in the direction either of over- or under-familiarity in respect of the immediate environment, or even of both simultaneously ("double" mental state). I cannot feel satisfied with any other explanation for these dreamy states as they occur in the course of epileptic processes (or for the reduplicative paramnesia of chronic epileptic and organic nervous disease) than that they represent dysfunction of the psychical mechanisms of perception and recognition in a plus or a minus direction, and that they are independent of any similarity or dissimilarity, contiguity, or other quality of the environment. This element of strangeness or unreality is very commonly met with in patients suffering from what may still be called psychasthenia. Janet ²⁴ has dealt with the condition, attributing it to diminution or absence of that cerebral activity which is characterized by complexity and richness of images, movements, and emotions. When the mind is accustomed to a certain maximum of consciousness, that maximum constitutes reality for the individual, and when it is not attained the mind no longer has a feeling of the present and of reality. It is interesting to note that Janet considers the phenomenon of *déjà vu* to depend on diminution of psychological tension, of synthetic activity, of consciousness of complexity of mental action and concentration. He too, therefore, though in a somewhat different way, links together states of hyper-familiarity and hypo-familiarity, as I may venture to call them. Considerations of this kind lead by easy stages to problems of depersonalization, which perhaps take us too far from our immediate subject. I may however cite the words of one of my psychasthenic patients.

"There seem to be two sorts of feelings in my thoughts, my own thoughts keeping me back, and other thoughts taking me away. When I go into the street I can see everything, of course, but things don't strike me in the old way. Formerly I knew that I was walking along ; now I don't seem to feel that I am. Things seem to be as in a dream. I can't fix myself when speaking ; I often go on talking to keep my end up, but I do not know that it is I who am talking. I feel as though I were not really there."

This patient is familiar with the *déjà vu* feeling, and has experienced it

frequently since he has been ill. "If I try to linger in the feeling, it always escapes if I direct my attention to it; but if my mind gets half-way, as it were, without being observed, I can always observe it."

We are brought back, nevertheless, to our problem by the following highly instructive case, illustrating the interrelation of epilepsy and psychasthenia.

CASE 13. H. M., male, age 24, had a typical epileptic attack ten months ago. Since then, he has suffered from "strange feelings" almost every day. They last for about half a minute. "When it occurs, everything round me, the objects, the people, seem strange; I can't understand them, they seem so strange. I always think, what are they here for? what are we walking about for? why am I doing what I am doing? At the same time, I always have a curious taste in my mouth, unpleasant and bitter, like bitter lemons. It comes always and only with the sensation."

In this case the reader will recognize the occurrence of a larval *folie de pourquoi* in the attack, identical with what is often found in psychasthenia, and he will not fail to note its significant association with the dreamy state and the crude sensation of taste.

If, finally, we endeavour to correlate transient disorder of psychical function, in the shape of mis-perception or mis-recognition, with what we may believe to be the physiological processes of epileptic conditions, we find ourselves unfortunately in the realm of pure speculation. On general grounds, we can assume that the mental states under review arise during activity of decontrolled centres, the "next lower" centres, as Jackson said. Psychical processes are not usually understood to be capable of focal localization, though the phenomena of aphasia do without question indicate that local lesions of the brain are often associated with partial disorders of a mental mechanism. In this connexion the views of some members of the Swiss school of neuropsychiatry deserve attention. Fankhauser²⁵ has elaborated in some detail an interesting conception, to the effect that the three outer layers of the cortex subserve ideation, the fourth is concerned with affectivity, and the inner two with perception. The suggestion is that the fourth is functionally linked with the outer three, forming the anatomo-physiological basis for psychical processes of the affective sphere accompanying the reception of sensory stimuli and the carrying out of ideational stimuli. Among these processes are included consciousness of self, apperception, and affect. In *perception* there is stimulation of sensory cortical fields (inner layers) and spread to the psychical layers (outer half), linked in the process

with an affective element ; in *ideation*, by association from memory is reproduced something resembling the idea of a perception ; in *illusion*, sensory excitations reach the sense fields, but for pathological reasons an inadequate innervation passes to the psychical outer half, and an imperfect idea of a perception results ; in *hallucination*, there is awakening of a sense area by a pathological idea (from the outer layers), or an idea accompanied by a pathological affect.

However schematic, artificial, and hypothetical such a conception may be thought, it does not lack support (von Monakow,²⁶ Berze,²⁷ Shaw Bolton²⁸). May not dysfunction as between outer (recognitive, ideational) and inner (perceptive) cortical layers, arising in the course of epileptic release, account for the phenomena of "fausse reconnaissance," illusion, spurious familiarity and unfamiliarity, "double consciousness," with which we have been occupied ? Did Miss E. C. (Case 6) make a lucky guess when she said, "My idea is that a layer of the brain gives way and another portion of it comes into being" ? Such a theory might account for the chronic reduplicative paramnesia of certain cases of organic nervous disease (general paralysis, senile dementia, chronic epilepsy) without doing violence to the facts ; we should then suppose that the abnormal passing of perception into recognition indicates dissociation of function of cortical layers, developing in conditions of diffuse cortical change and showing itself for a longer or shorter period in accordance with the anatomo-physiological circumstances accompanying its appearance. As an aura to, or part of the semiology arising along with, an epileptic process, it would be equally conceivable that perception should be transformed erroneously into recognition, or that recognition should be stripped of its qualities and be erroneously judged to be mere perception, because of such dissociation of normally integrated cortical function.

And is there nothing in the clinical observation, many times made, that this dysfunction is especially prone to develop along with crude olfactory and gustatory sensations whose basis is, precisely, excitation of the uncinate region of the temporal cortex ? The combination passes outside the range of pure coincidence. Similar associations are not in my experience noted in respect of auditory or visual auræ, although either or both of these may form a constituent if transient feature of dreamy states. Evidently they are not anything like so potent in arousing the psychical phenomena of the states, but merely in their turn are stimulated or released into action as the epileptic process sweeps

over the brain from its uncinate nodus. To this matter I shall return at the close.

The "Panoramic Memory" Type

The last point to which attention may be directed is the "memory" type of aura. Its occurrence has been remarked often enough in abnormal conditions other than epilepsy. Most people are aware of stories of rescue from drowning where the person concerned, losing consciousness as a result of immersion, has nevertheless afterwards declared that during that stage all the events of his past life have chased each other through his mind as one panoramic picture succeeds another. Such cases are well authenticated. And it is important to note that the actual time occupied by this astonishing survey of the individual's previous existence has never been longer, objectively, than a minute or two.

I may cite the personal experience of Admiral Sir Francis Beaufort.²⁹

"Many years ago, when I was a youngster on board one of His Majesty's ships, in Portsmouth harbour, after sculling about in a very small boat, I was endeavouring to fasten her alongside the ship to one of the scuttle-rings; in foolish eagerness I stepped upon the gunwale, the boat of course upset and I fell into the water, and not knowing how to swim, all my efforts to lay hold either of the boat or of the floating sculls were fruitless. I was soon exhausted by my struggles and before any relief reached me I had sunk below the surface.

"From the moment that all exertion had ceased a calm feeling of the most perfect tranquillity superseded the previous tumultuous sensations. My sensations were now rather of a pleasurable cast, partaking of that dull but contented sort of feeling which precedes the sleep produced by fatigue. Though the senses were thus deadened, not so the mind; its activity seemed to be invigorated in a ratio which defies all description, for thought rose after thought with a rapidity of succession that is not only indescribable, but probably inconceivable, by anyone who has not himself been in a similar situation. The event which had just occurred, the effect which it would have on a most affectionate father, and a thousand other circumstances minutely associated with home, were the first series of reflections that occurred. They then took a wider range—our last cruise—a former voyage—my school—and even all my boyish pursuits and adventures. Thus travelling backwards, every past incident of my life seemed to glance across my recollection in retrograde succession, not, however, in mere outline, but the picture filled up with every minute and collateral feature; in short, the whole period of my existence seemed to be placed before me in a kind of panoramic review; indeed, many trifling events which had been long forgotten then crowded into my imagination, and with the character of recent familiarity.

"The length of time that was occupied by this deluge of ideas, or rather

the shortness of time into which they were condensed, I cannot now state with precision, yet certainly two minutes could not have elapsed from the moment of suffocation to that of my being hauled up."

This graphic narrative reveals in perfection the features characteristic of the panoramic memory aura ; we may note particularly how old, far-off events were tinged with the feeling of recent familiarity. The late Canon Liddon ³⁰ referred to an analogous experience in a famous sermon entitled "The First Five Minutes after Death."

"It once happened to me to assist at the recovery of a man who nearly forfeited life while bathing. He had sunk the last time and there was difficulty in getting him to land, and, when he was landed, still greater difficulty in restoring him. In describing his experience of what must have been the whole conscious side of the act of dying by drowning, he said that the time had seemed to him of very great duration ; he had lost his standard of the worth of time. He had lived his whole past over again ; he had not epitomized it ; he had repeated it, as it seemed to him, in detail and with the greatest deliberation. He had great difficulty in understanding that he had only been in the water for a few minutes. During these intenser moments of existence the life of the soul has no sort of relation to what we call time."

Such remarkable happenings, however, are not confined to those who have nearly lost their lives by drowning. A number of illustrations of this are collected in Forbes Winslow's ³¹ forgotten book entitled *Obscure Diseases of the Brain and Mind*, and one may be quoted.

"A gentleman, during an attack of acute mental depression, hanged himself. He often related to me the strange mental visions that floated in his mind during the few minutes or (in all probability) seconds he continued suspended, and temporarily deprived of consciousness. They were of the most pleasing character. The scenes of his early life were, in their minutest particulars, revived. Incidents connected with the school in which he received his early instruction were reproduced to his mind. The remembrance of faces (known when a child) that had been (as he supposed) entirely obliterated from his memory, was restored to his recollection in a most remarkably truthful and vivid manner. During that critical second of time (when it may be reasonably presumed he was struggling with death) every trifling and minute circumstance connected with his past life was presented to his mind like so many pictorial sketches and drawings."

An impressive instance of the same phenomenon has occurred in my own experience. A friend of mine choked at the dinner-table, and rose to leave the room. He reached the hall and there sank quietly in a semi-asphyxiated condition to the floor. His father had noted the incident, and his quick ears had detected the

fall. He rushed out of the room, saw his son's urgent condition, and without hesitation crooked his little finger into his son's throat and withdrew the piece of meat impacted in the upper larynx. The time that elapsed was at the most one minute. Yet during that brief period of unconsciousness my friend had gone back in thought, in the most pleasant way in the world, to scenes and incidents of his earliest childhood. Trifling events of his nursery life, his childhood and school days, passed before him in a succession of vivid pictures. He saw again the furniture of his nursery, its paper and carpet, his nurse, and so on, and he has told me how astounded he afterwards was to think that these things could all thus rise in clear outline before him.

At moments of intense danger, when the senses are benumbed by terror, similar eternities have been lived through. The following excerpt is taken from an officer's letter, published in *The Times* newspaper at an early period of the War.

"We lay there in the potato crop like partridges. I think we were all too petrified to move. We lay under that shell-fire for three hours, and I think that none of us will ever forget the feeling of thinking that the next moment we might be dead. I kept wondering what it was going to feel like to be dead, and all sorts of little things that I had done and places I had been to years ago and had quite forgotten kept passing through my mind. I have often heard of this happening to a drowning man, but have never experienced it before and don't want to again! My brain seemed extraordinarily cool and collected, but I looked at my hands and saw them moving and twisting in an extraordinary way, as if they didn't belong to me."

No one can fail to notice what these diverse cases possess in common. It is the definite resuscitation of an apparently unending series of mental pictures, back to infancy, released during partial asphyxiation from drowning, hanging, or choking, or during a state of emotion sufficiently intense to paralyse the faculties and overwhelm cortical control. And we have seen that identically the same phenomenon may be experienced as part of an epileptic process, commonly associated with hallucinations of taste or smell.

The conclusion seems incontrovertible, that when full consciousness is in abeyance previously organized mental states rise into a secondary consciousness; oblivion of actual environment is followed by heightened activity of visual and other sense elements, constituting a dream-like consciousness in which space and time are obliterated; it is in a scarcely metaphorical sense "another life." Of immediate interest to the student of the subject is the allied

question : If the phenomenon is caused merely by decontrol, why does it not occur more often—as far as epilepsy is concerned, much more often ? To this question several considerations are apposite.

Rapidity of development of epileptic processes may effectually prevent any intermediate stage of double consciousness from establishing itself. Its momentary appearance may be drowned by a brutal sweep of the discharge over the cortex as a whole. Obviously, a certain relative slowness is essential if the aura is not to be submerged in the mental nothingness of complete conscious loss. In the second place, constitutional differences between visuals and auditives may here obtain, though I do not stress the point. The cortical disposition of the individual concerned should not be ignored, since visuopsychic mechanisms may in him be relatively poorly developed, and therefore incapable of being energized into abnormal functional action. Again, we must give due weight to psychological qualities associating smell or taste with vision, and to anatomical relationships between temporal and occipito-parietal areas of the cortex. That a taste or smell, particularly the latter, exercises a remarkably potent influence in awakening visual and other memories is a psychological commonplace and is known to every student of literature. I must content myself with a solitary citation from *The Private Papers of Henry Ryecroft*.³²

“ A little plant of which I am very fond is the rest-harrow, which sometimes grows in sandy ground above the seashore. In my childhood I have many a time lain in such a spot under the glowing sky, and, though I scarce thought of it, perceived the odour of the little rose-pink flower when it touched my face. Now I have but to smell it, and those hours come back again. I see the shore of Cumberland, running north to St. Bee's Head ; on the sea horizon a faint shape which is the Isle of Man ; inland, the mountains, which for me at that time guarded a region of unknown wonder. Ah, how long ago ! ”

Accepting, then, the psychological alliance between these two senses, we may ask whether it is not also based on anatomo-physiological connexions. The inferior longitudinal fasciculus is, next to the corpus callosum, the largest bundle of association-fibres in the whole brain. It unites temporal and occipital cortex, ensuring a wealth of reciprocal interaction. While odours often arouse visual pictures, the reverse occurs with far less frequency ; anyone, almost, can summon up visual memories, but few indeed are capable of doing the same with smells or tastes. Hence one may

suppose that while uncinate excitation in gustatory or olfactory cortex is likely to be succeeded by visual excitation, reversal of the sequence is improbable. This is in accordance with clinical experience; uncinate aura is frequently followed by visual memory complex; the latter, forming the aura, much less often awakens in its turn uncinate elements.

We may take it, therefore, that dreamy states of the different varieties here illustrated and analysed, occurring as preliminaries to epileptic convulsions, indicate the development of a process commencing in all probability in the temporal lobes and spreading therefrom by anatomo-physiological lines. Epileptic discharge may be responsible for crude hallucinations of smell and taste, epileptic decontrol for the development of mental states of the kinds discussed. Possibly epileptic spread tends to be slower along temporo-occipital than along fronto-rolandic pathways, hence there is more time for still normal mechanisms to rise into released activity in the case of the former than in that of the latter. Temporal epilepsy exhibits special features of its own, and its psychical components present close resemblances to phenomena which we have seen develop in normal persons under abnormal circumstances. I do not think it illegitimate to assume a decontrol in the latter instances affecting such cortical regions and mechanisms as are decontrolled when the epileptic process starts its spread from the uncinate sphere. Thus clinical study illuminates normal cerebral function on both physiological and psychical sides, however much of the complex problem presented by this special variety of epilepsy still remains for full solution.

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CHAPTER V

THE NARCOLEPSIES *

Introduction. Nosology. Terminology. Personal Cases. Classification and Clinical Varieties. Clinical Symptoms: Sleep; Attacks of Loss of Tone and Power; Transitions and Substitutions. Relation of Narcolepsy to other Clinical Syndromes: Myoplegia; Catalepsy; Epilepsy; Oppenheim's *Lachschlag*. Etiology. Pathology. Sleep Centres and their Localization. Pathogenesis. Conclusions.

Introduction

Influenced largely though not solely by the world-spread of epidemic encephalitis since 1918, neurology has in recent years turned its attention in increasing measure to problems presented by disorders of the function of sleep. Among these is the condition called narcolepsy—a term that has always been loosely employed, various writers having described clinical examples of sleep disorder under this heading without staying to define the sense in which it is being used. As a consequence, current ideas are uncommonly fluctuating where it is concerned, though, as we shall see, attempts at delimitation have recently been made. A legitimate matter for discussion is whether, in point of fact, clinical instances of disorder of sleep are now occurring with greater frequency than in other times. This is evidently the opinion of some recent writers, yet long before epidemic encephalitis appeared many investigators had interested themselves in narcolepsy and cognate problems of sleep, as anyone who makes himself familiar with the older literature (which is far indeed from being scanty) can readily realize.

My own experience in this respect may be given a passing allusion. I was house physician to the late Sir William Gowers at the time (1907) he wrote his fine little volume on *The Borderland of Epilepsy*,

* A paper read at the Annual Congress of the Association of Physicians, Belfast, June 3, 1927, and reprinted from *Brain*, 1928, vol. li, p. 63. The references to all the writers mentioned in the text will be found at the close of the chapter.

included in which is a chapter devoted to some sleep symptoms—among them, narcolepsy. I became familiar at that time with the subject, but in twenty years saw no definite example of narcolepsy (with one or two minor exceptions); yet in the course of the last year I have met with some half-dozen cases, the details of four of which are duly recounted below. Spiller (1926) appears to have had an analogous experience: “A few years ago it was exceptional for a neurologist even with a large experience to see a case of typical narcolepsy. I have been impressed by the fact that at least three cases have come under my observation within a year.” From these considerations a case might be made out for an actual increase in disordered conditions of sleep somehow related to the times in which we are living, and their ascription to processes arising out of epidemic encephalitis might be plausibly offered as one explanation, but at present I am far from being convinced of any frequent etiological association of the latter with the former. Whether any apparent increase can be accounted for, to some extent, by augmented attention on the part of clinicians is a speculation not easily to be substantiated one way or the other; and as for the actual question of numbers of cases it also cannot be decided in any positive sense.

Nosology

From the viewpoint of nosology it is unscientific to allude to any of the disorders of sleep as *diseases*. In particular, to describe the variety called narcolepsy as a *morbus sui generis* is a nosological error, yet in the communications of Henneberg, Redlich, and others this regrettably occurs *passim*. That a special *clinical* type of sleep disorder, a clinical syndrome corresponding to Gelineau's original description of a “neurosis” to which he gave the name “narcolepsy,” exists, may or may not be regarded as established; but, at the most, it remains only a clinical combination. There can be no narcolepsy of any kind without a cause. No one has put the matter more clearly than Lhermitte,⁴⁵ as the following citation shows: “Admettre la nature névrosique de la narcolepsie, c'est admettre par le fait même qu'elle est une maladie au vrai sens du mot, c'est-à-dire un état pathologique lié à la même cause et ressortissant à la même thérapeutique. L'énoncé même des caractères essentiels d'une ‘maladie’ suffit à montrer que la narcolepsie ne peut à aucun titre être assimilée à une affection autonome.” And even if we take the particular syndrome of

narcolepsy in the sense understood by Gélinau, in more than one recent case it has appeared as a sequel to epidemic encephalitis, rendering its isolation as a *morbus sui generis* more impracticable even than before.

Terminology

As already remarked, the term "narcolepsy" has been employed for more than forty years (Gélinau's paper having been published in 1880) in the vaguest way to signify any clinical variety of paroxysmal diurnal sleep, in recurring attacks of variable duration; or, indeed, of prolonged sleep-states, the association of which with such known conditions as hysteria or other psychoneurosis or psychosis is unmistakable. Gélinau's definition—"Une névrose rare, caractérisée par un besoin subit, irrésistible, de dormir, ordinairement de courte durée, se produisant à des intervalles plus ou moins rapprochés et obligeant le sujet à tomber ou à s'étendre pour lui obéir"—is not of itself calculated to delimit the conception. Gowers was one of the first to desiderate its restriction, though present-day writers have neglected his work; he pointed out that cases of *continuous* sleep should have another designation, as also those in which a sleep-state is interrupted only when the sufferer is roused, for which he rather favoured the discarded term "somnia." His conclusion was that narcolepsy should be used solely for cases in which "definite brief sleep interrupts a normal state." Naturally enough, it is impossible to say where the precise line is to be drawn between short and prolonged sleep (useful though the distinction is), and, for that matter, between sleep and lethargy or trance; a recurrent tendency often characterizes trance-like states, while the question of whether the patient wakes spontaneously or only if roused cannot be utilized as a diagnostic criterion, since both conditions are frequently seen in one and the same case at different times. The truth is that all these distinctions are highly artificial, and the term ought to be used solely in a symptomatic sense, to include all states of recurring diurnal sleep, whatever their etiology.

A tendency, however, has revealed itself since the publication of Loewenfeld's paper in 1902 to regard the narcolepsy of Gélinau as a special form constituted by a double symptomatology, viz. the combination of attacks of sleep with transient attacks of physical powerlessness under the influence of emotion, so that the patient's knees give way and he may sink to the ground, without any loss of

consciousness ("cataplexy"). This interesting phenomenon was noted by Gélinau's patient to occur when he laughed heartily, was observed also in Loewenfeld's case, and was raised by the latter author to the level of a diagnostic feature of narcolepsy *sensu strictiori*. Following him, Henneberg, Redlich,⁶⁸ Stöcker, Kahler, Jolly, Singer, Münzer, and some other German authors consider the combination pathognomonic of narcolepsy, and would reserve the use of the term for such cases only, a line which is also strongly advocated by Adie in his recent able thesis on the subject. On the other hand, Goldflam, Curschmann and Prange, and others hesitate to regard attacks of powerlessness as essential to the conception; Matzdorff is convinced of the occurrence of rudimentary and atypical cases which do not correspond in toto to the above-mentioned combined form, and in this respect is followed by Stiefler⁸⁷ in his latest paper. Along with a typical case two others characterized only by sleep attacks are included by Janzen under the same designation, while, as it happens, a similar attitude is adopted by Adie, who accepts a case of his own, and two reported by Goldflam, in which no powerless phenomena were observed, as belonging to "true narcolepsy," and says in so many words that "sleep attacks alone may occur in genuine cases."

Experience should surely have convinced the neurologist that no good purpose is served by hard-and-fast schematization of a clinical syndrome to fit a conception based solely on an original description; although the four personal cases detailed below belong clearly to the combined category of sleep plus emotional ("cataplectic") attacks, my contention is that this category is nothing else than a special variety of narcolepsy, and that the latter expression can have no more than a purely semiological significance.

This consideration leads to a further point in terminology. The Greek word *νάρκη* signifies, according to Liddell and Scott, "numbness, deadness (Lat., torpor), caused by palsy, frost, fright, etc.," whereas the characteristic of the clinical states that here concern us is their *apparent* resemblance to ordinary sleep. Hence the alternative term "hypnolepsy" has been suggested, not originally by Singer (as he, in ignorance of previous work, appears to think), but by Foot, as long ago as 1886. It was also advocated by Carl Camp twenty years ago (1907). But "narcolepsy" has been given so long a start that perhaps it will now scarcely be overtaken.

The most recent terminological suggestions come from Wender-

owic, who proposes "hypnolepsia genuina" for the Gelineau combined type, "hypnolepsia symptomatica" for the same type appearing as a sequel to, or in association with, a known pathological condition, and "narcolepsia" for symptomatic hypnoid conditions arising in the course of various diseases. I do not feel inclined to adopt this unnecessarily dichotomizing classification, and conclude by repeating that for present purposes "the narcolepsies" stand for hypersomnic varieties of sleep disorder, whatever their etiology, among which it is convenient to distinguish a combined type corresponding to that so well differentiated by Gelineau.

Personal Cases

CASE 1. *Male case. Duration, three years. Cause unknown. Typical diurnal seizures of "natural" sleep of brief period, associated with typical attacks of tonelessness in limbs under the influence of emotion. Additional symptom, frequent attacks of diplopia.*

P. O. R., age 24. Occupation, railway carpenter, unmarried.

History of present illness.—In April, 1924, the patient had his first attack of irresistible sleep when actually on the top of a signal-post executing a repair. He managed to climb down and fell fast asleep at the foot of it, for perhaps half an hour. Ever since, he has suffered from continually recurring diurnal attacks of sleep, not a single day passing without them. About the same time as the original attack, or rather later, he noticed occasional double vision, independent of the sleep turns, the images being on the same horizontal level and a little way apart. At a period subsequent to April, 1924, but not determined with exactitude, he noticed that whenever he laughed heartily he suddenly experienced a giving way of the knees, so much so that on several occasions he sank to the ground powerless, but was up again a few seconds later. Apart from this triad of symptoms he has no other complaint, and has continued at his work.

Previous history.—Two years previously, playing football, he met with an accident and was badly concussed, but the skull was not fractured. Venereal disease has never been contracted.

Family history.—Nil ad rem.

Condition on examination.—The nervous system is normal in all respects. The patient has not put on weight, weighing at present 12 st. 4 lb., the same as at the outset. His facies reveals a state of mind of some anxiety in respect of emotional influences; rather frowning and somewhat subdued, he preserves a forced gravity. "I don't laugh any more than I can help" (see Fig. 2).

Wassermann test in the serum, negative. X-ray picture of sella turcica and vicinity, normal. Blood-sugar estimation (Dr. O'Flynn), normal curve.

This case conforms entirely to the syndrome of the first division of the first group (see below), presenting the combination of attacks of sleep and of cataplexy. The former are always of brief

duration, rarely exceeding ten minutes. From them the patient either awakes spontaneously, or can be at once aroused by a touch ; he immediately feels fresh and continues his job. There is no background of continuous sleepiness, and no conscious feeling of physical tiredness or fatigue. The patient has slept many times while waiting his turn to come into my out-patient clinic, yet never once he has entered, however long the examination. Personal observation of the sleep shows it to be seemingly a state of natural slumber, with no abnormal manifestation. At night, he passes

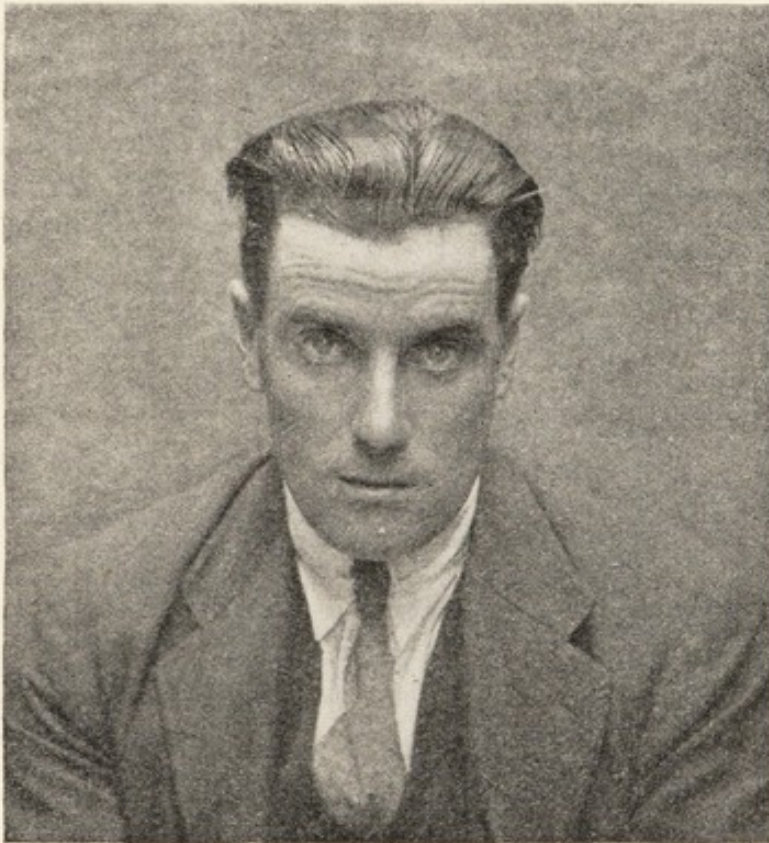


FIG. 2.—Facies, Case 1.

some eight hours normally in sleep and has no difficulty in awaking. In regard to the “powerless” phenomena, the sole excitant appears to be laughing, no other modality of emotional stimulus precipitating them.

CASE 2.—*Male case. Duration, nineteen months. Cause unknown. A brother suffers from some variety of epilepsy. Typical diurnal attacks of sleep, and numerous attacks of cataplexy, especially with laughing but also under the influence of other emotions. If the patient forcibly keeps himself awake an attack of powerlessness will supervene instead of the sleep. Neurological examination during one of these*

reveals temporary complete loss of deep reflexes and an extensor response. Additional symptom, increase of weight, amounting to 2½ st. since onset.

E. C., age 26. Occupation, bricklayer; married.

History of present illness.—One Saturday evening, nineteen months ago, the patient was handed a paper informing him that he had won £5 in a football competition sweepstake. At this unexpected news, in the very moment of excited rejoicing, he suddenly felt helpless, his head dropped down, his limbs gave way under him and he “flopped” to the ground in a state of complete powerlessness. His head was sunk on his chest and he remained motionless, unable to move a finger, for about a minute. During the whole of the time he was conscious of what was going on round about him. Immediately on recovery he felt perfectly well.

A short time afterwards (exact interval not known) he had his first attack of sleep, the circumstances of which are not remembered.

Ever since, he has had attacks both of sleep and of loss of power in the limbs under the influence of any emotion, the former occurring every day, often many times a day.

(1) *Sleep attacks.*—These are apt to occur particularly during his work, that of bricklaying. He often finds it monotonous, laying one brick after another, and the attack is usually ushered in by his mind wandering off his job: “I feel so used to it that my mind wanders off, where, I cannot tell, and then I drop off.” The duration of the sleep is from three to four minutes; it is not preceded by any feeling of physical tiredness, but rather by this sort of “day-dreaming.” Whenever he awakes he feels quite fresh. He either awakes spontaneously or is easily roused by a touch or a shake, but during the attacks he is soundly asleep, oblivious of his surroundings, and as a rule not dreaming. He has often fallen asleep at meals, “with my fork half-way to my mouth.” On the whole, he is as liable to fall asleep in the evenings at home as at work, yet he is confident that the warmth of the room, the time and place, make no difference, nor does the matter of interestedness arise. “I fall asleep at the pictures, or during a business conversation, just the same.”

(2) *Attacks of loss of power and tone.*—The attacks of “tonelessness” arise in two distinct ways, either under the influence of emotion, or *because he is trying to avoid an oncoming sleep attack.* In regard to the first of these, any excitement suffices. Thus: “at a football match my legs give way just when some one is about to score a goal.” Temper will bring on an attack, and the tendency is always aggravated if the patient thinks that people are looking at him. “If I am playing the fool with the baby and my wife looks on and ‘chips in,’ I am sure to have an attack.” Laughing heartily never fails to bring on one of these turns. It is noteworthy that there is sometimes a relation between the emotion and the part of the body involved for the moment in the particular action concerned. Thus: “If I go to throw a stone at a glass bottle and any of the fellows are looking, the power seems to go out of my arm.” “If I go to shoot the ball at goal and I see the fellows watching me, the use goes right out of my leg; I simply can’t do it.”

According to the patient’s own description, in these seizures the head falls

forward on the chest in a nodding movement, the limbs are absolutely useless and powerless, the whole thing lasts a minute or two, and immediately after he is "as right as rain" and has no desire to sleep for the next hour.

As for the second causation, he has for long observed that *if he endeavours to keep himself awake forcibly, when sleep is imminent, an attack of powerlessness will supervene*, unaccompanied by any conscious emotion (unless we are to assume that the effort to avoid sleep is itself coloured with emotional tone). In the event, one or other attack is certain, though there have been occasions when deliberate shrugging of the shoulders has seemed to prevent it.

Of the greatest interest is the fact that when he has been asleep and dream-



FIG. 3.—Facies, Case 2.

ing, *the emotional content of the dream has precipitated an attack of powerlessness*. The worst he has ever experienced lasted for about a quarter of an hour, and occurred under the following circumstances. He had fallen asleep and was dreaming of a murder. The emotion experienced in association therewith brought on one of the "loss of power" attacks, so that he at once awoke and was fully conscious but was utterly unable to move a single finger. He thought he heard his brother and sister coming up the stairs, he tried to call to them but could not make a sound; the more he tried the more intense became his emotion and the more absolute his helplessness; he lay thus, flat on the floor, motionless but suffering acute mental distress, for some fifteen minutes ere the attack dissolved itself spontaneously. Similar

but less severe attacks have occurred when he has been dreaming of "terrible happenings" on other occasions.

Ordinarily, the patient's night sleep is of normal duration, and he wakes readily enough in the mornings.

At the commencement of his illness—which has never interfered with his daily work—he weighed 10 st. 6 lb.; he now weighs 13 st. (see Fig. 3).

Previous history.—He has had no previous illness of any import, and no head injury.

Family history.—An elder brother, age 32, suffers from some kind of fit. These began in 1919 and he has had only three in all. From the description given it is difficult to determine their exact nature; it appears he falls unconscious and is stiff in his limbs, but there is no change of colour, tongue-biting, or involuntary micturition. As he served in the army during the War and saw much fighting, it is possible they have a psychogenic basis. Otherwise, the family history is negative.

Condition on examination.—The nervous system is normal in all respects. Wassermann reaction in the serum, negative. X-ray examination of the sella turcica shows it to be if anything on the small side. Blood-sugar curve (Dr. Lawrence), normal in character.

The clinical value of this impressive case of combined narcoleptic and emotional attacks is enhanced by the fact that I was able to observe one of the latter from beginning to end and to examine the patient's neurological condition during it. Dr. MacDonald Critchley, Registrar at the National Hospital, was with me at the time and helped materially in the examination, which was of necessity hurried.

Description of cataplectic attack.—While I was occupied with another patient, E. C. was sitting behind me and, as afterwards appeared, was endeavouring to keep himself from falling asleep. Suddenly I heard a slight groaning sound coming from him, and at once looking round I saw his head nodding gently on his chest as he sank forward into a bent position and a moment or two later slid or "slithered" off the chair to the floor. The arms were by the side and the fingers semiflexed; the eyes were closed. Lifting the arms I found them absolutely atonic and flaccid; when let go they fell like lumps of lead by the side. The legs similarly were completely atonic, sprawling out on the floor. Lifting the eyelids I observed that the pupils reacted to light slightly but definitely, while the eyelid muscles were so toneless that little or no reflex contraction took place on my touching the cornea. Testing the knee-jerks I found them *completely abolished* on both sides. In the meantime Dr. Critchley had hurriedly taken off the patient's shoe and sock on the left side, and testing the plantar reflex obtained a slight but left definite *extensor response*, which he demonstrated to me and which I corroborated.

Just as we were finishing this rapid examination the patient suddenly said, "I'm all right, sir," in his ordinary voice, and with a faint smile. Muscular power came back; he moved his limbs, got on to the chair, and told

us he had been conscious the whole time. He described what we had done, from lifting the eyelids to scratching the sole of the left foot. The duration of the whole attack was from sixty to ninety seconds. Testing his knee-jerks again, I found them *active*, even brisk, on both sides, two minutes after he had recovered.

So far as I am aware this is the first time a neurological examination of a patient in the cataplectic state has been recorded. When Adie's paper was published (September, 1926) it was therein specifically remarked that "no patient has been examined by a competent neurologist while in the cataplectic state."

I shall refer at a later stage to the significance of this observation, supplying as it does details of much importance for the study of the mechanisms involved and for the problem of the nature of the emotional attacks and their relation to the narcoleptic phenomena.

CASE 3.—*Male case. Duration, twenty years. Cause, unknown. There are three separable types of attack in this case: (1) typical short diurnal sleep seizures; (2) typical (momentary) attacks of loss of power in the limbs under the influence of any excitement, laughter or rage; (3) trance-like states of "absolute paralysis" during which no movement is possible, while consciousness is intact, brought on not by emotion but usually when patient is resting or sitting quietly, sometimes when on his feet.*

T. T., age 41. Occupation, boilerman; married.

History of present illness.—(1) As far as the patient is able to recall, he first noticed the tendency to *attacks of sleep* when he was in the Army in India, some twenty years ago. Scarcely a day has passed since then without his having fallen asleep, not once but a number of times. Without any preliminary warning in the shape of tiredness or sleepiness he suddenly falls "dead asleep"—a dreamless slumber of some four or five minutes' duration from which he wakes spontaneously, or is aroused with ease and promptness. He has had "dozens" in the day. He has fallen asleep on his feet, and on one occasion injured himself by knocking against a lamp-post. He has had such attacks at his work, during meals, under every variety of circumstance, and cannot assign them to any special cause, such as monotony of occupation, warmth, time of year, diversion of interest, etc. As soon as the sleep is over he feels fresh and well, and makes no complaint of physical fatigue or of any general sleepiness.

(2) In addition, and for about the same period, the patient has noticed that any *emotional excitement causes a peculiar sensation of weakness in the limbs*, which results in their giving way so that on numerous occasions he has sunk down on his knees, or actually fallen. For instance, on one occasion he saw a horse bolt and without further warning his knees weakened under him and he went down. Its duration is, however, exceedingly brief, "almost

momentary," and yet there is time for him to be well aware of his heart "beating like anything," and of a definite feeling of tingling over the head, cheeks, and body, "like a lot of strings moving over my face." "For the moment I feel as if my blood were clotting." Laughter brings on these attacks of loss of muscular power, as well as annoyance or anger. In his own words: "If I were to try to strike one of my children it would come straight over me before I could do so, and I would go flat on the floor." This has actually happened on more than one occasion.



FIG. 4.—Facies, Case 3.

(3) The third type of attack is as follows: At any time, usually when sitting quietly in a chair, lying on a sofa, leaning against a wall, and so on, there will suddenly steal over him a "paralysing feeling"; his eyes close but he never loses consciousness and is acutely aware of what is going on, his limbs relax, and while the attack lasts he is *absolutely incapable of moving* a finger-tip through a fraction of an inch. "I am paralysed from head to foot; some one could murder me and I could not move." On more than one occasion he has had a cigarette in his hand when this type of attack came on, and has actually burnt his fingers badly without being able to make a single movement to let it drop. The patient says that he "fights against it" inwardly by every means he can think of, yet not a sign of this is visible to the looker-on, who sees him lying or sitting motionless. It appears that

he eventually is able to make a grunting noise in his throat to attract the attention of his wife or other person; if he is touched or shaken forcibly the spell is at once broken and he recovers instantaneously. During the attack he is unaware of any feeling of fear, though he can neither move nor speak, but he is frequently conscious of his heart beating, "as if it were bursting." The duration of the attack is usually from three to five minutes.

He made spontaneously the interesting statement that "if I was to try and prevent myself from a sleeping attack one of my paralysing attacks would come on."

The patient's ailment has never prevented him from continuing at his work, and he does not complain of anything else than the symptoms described above. For years he has given up all thought of amelioration, having consulted many doctors and visited various hospitals without avail.

Previous history.—Rheumatic fever at the age of 9. Extremely moderate in alcohol. Venereal disease denied. No accidents.

Family history.—One of his five children died with rheumatic fever. Otherwise, negative.

Condition on examination.—The nervous system is normal in all respects. Has not put on weight to any extent (see Fig. 4). X-ray of sellar region shows it to be normal. Wassermann test in the serum, negative. Heart normal. Blood-pressure, 135 mm. Blood-sugar normal, and complete urinalysis also (Dr. Lawrence).

This third personal case similarly conforms to the group of combined narcoleptic and tonelessness attacks, and is of especial value in that it furnishes evidence of the occurrence of trance-like states independent of emotional excitation, and of a transition from sleep attacks to "paralysing" attacks if the former are forcibly checked. Cases 2 and 3 supply clinical data which throw light on the possible interrelation of the sleep and the emotional syndrome. To these matters we shall return immediately.

CASE 4.—*Male case. Duration, two years. Cause, indefinite illness two or three months before, diagnosed as epidemic encephalitis. Typical short diurnal attacks of sleep, with attacks also of loss of power in the limbs (often local) under the influence of emotional stimuli.*

A. B. C. D., age 18. Schoolboy.

History of present illness.—Nearly two and a half years ago the patient, when at school, suffered from some kind of febrile ailment, thought at first to be influenza but later asserted to have been epidemic encephalitis. No diplopia was complained of, nor was abnormal sleepiness or sleeplessness a symptom. Within a period of not more than a month or two, however, he put on no less than a stone and a half in weight, and at the same time began to have sudden attacks of "sleep" at meals, or when unoccupied or uninterested. An imperious desire to close the eyes was the aura, and immediately they closed he fell asleep. The duration was usually extremely brief,

amounting merely to half a minute or even less, but on occasion it extended to a quarter of an hour. They have continued without intermission ever since, recurring several, sometimes many, times a day. In these attacks the patient is not as a fact deeply asleep; on the contrary, he is immediately aware when he is touched, when he is spoken to, when anything goes on round him; on a number of occasions he declares he has not been asleep at all, but has presented merely the appearance of sleep, for his state, he thinks, is more akin to that of "day-dreaming," a sort of "reverie" which is different from ordinary sleep. Once, in a railway train, the desire to close the eyes overmastered him, and instantaneously a book he was holding fell heavily from his hands; the abruptness of the onset of the sleep attacks always strikes him as peculiar. Night sleep is normal and of normal duration.

From the commencement of the affection another, to him very curious, symptom has manifested itself. It showed itself first on the occasion of a school steeplechase. He had to jump a hedge, and as he approached it running he suddenly felt a momentary anxiety whether he could do it or not. On the instant his limbs relaxed and he fell helpless against the hedge, completely unable to get his legs to answer his call for an effort. Within half a minute he had recovered and surmounted the obstacle without difficulty. On many occasions since, a similar but very brief failure of power has made its appearance, notably when he is playing tennis. Just as he is about to serve the ball an anxious thought flashes through the mind that he cannot do it and an equally momentary weakness of the right arm develops; by an effort he overcomes the loss of power and delivers his service usually so well that the momentary hesitation is scarcely observed.

The toneless or powerless attacks are thus largely local and of extreme brevity, but it is otherwise with the act of laughter. He has cultivated what he calls an "artificial" or "unnatural" laugh, because on one occasion when laughing heartily he became alarmed at a sudden weakness in both his legs, so that he nearly fell to the ground. As there were onlookers he felt highly sensitive about it; he now avoids the possibility of a recurrence by laughing in an unnatural way, "outwardly" and not "inwardly." He has some difficulty in describing exactly what he does at such times, but it is sufficient to prevent development of the phenomenon of temporary weakness.

Previous history.—With the exception of the illness already alluded to (? encephalitis) the previous history is negative, and there is no record of head injury.

Family history.—On both sides of the family cases of "bad temper" in an aggravated form have occurred. His mother is an "exceptionally nervous woman."

Condition on examination.—A bright and highly intelligent boy, with a fine scholastic record, he shows no sign of any organic affection of the nervous system. All the usual methods of examination have proved consistently negative.

The clinical interest of Case 4 resides mainly in the inapplicability of the term sleep to the first kind of seizures, which more closely resemble reveries (though outwardly they are like the sleep state); in their absolute suddenness and brief duration; and further, in

the almost larval or embryonic type of the cataplectic attacks, which are often scarcely more than momentary and may well pass unobserved ; in their frequent restriction of range, and in the apparent success with which the patient manages to prevent them from reaching fuller development.

Classification

In the present state of knowledge a complete etiological or pathological classification is out of the question, and we are compelled to fall back on an empirical clinical grouping. A purely tentative division along the following lines may prove clinically convenient.

I. Recurring diurnal attacks of sleep : (a) associated with attacks of "tonelessness" ; (b) without "toneless" attacks.

II. Sleep attacks of prolonged duration, day and night sleeps running into each other ; continuous sleep.

For various clinical examples of these hypersomnic conditions a pathological basis has been found, but it has not yet been discovered for all. The range of the pathological substratum is so extensive, covering cerebral tumours on the one hand, and psychopathological states on the other, that a classification founded on it would not provide any clearer idea of the essence of the disorders.

Taking the above empirical grouping, we may consider it in detail.

Clinical Varieties

I. Cases which definitely belong to the first subdivision of the first class (sleep attacks with attacks of "tonelessness") must still be regarded as relatively and absolutely rare. In his monograph Adie has given a *précis* of those (in males) reported by Gélineau, Henneberg, Goldflam, Redlich,⁷⁰ Jolly, K. Mendel, Noack, Somer (2), Loewenfeld, Stiefler,⁸⁵ Stöcker (his Case 4), and Singer, and has added one male case of his own—a total of 14, to which he appends 3 personal female cases—17 in all. Typical cases (in males) published before his own communication, but not included by him, are those of Curschmann and Prange (their Case 2), Westphal, Carl Camp, Redlich⁶⁹ (2), Wenderowicz, Matzdorff, and Bruno Fischer—8 more ; since his paper appeared there have been added those of Janzen (his Case 2), Hilpert, Spiller (3), and the first three here supplied by myself—a total of 30 male cases. In respect of this first variety of narcolepsy in the female sex, Adie says that "hither-

to the only published female cases were two described by Nevermann and by Kollewijn of "narcolepsy in pregnancy," but while these are judged by him not to have been cases of "true narcolepsy," "for sleep attacks alone occurred and they ceased after delivery," he has overlooked those recorded by Kahler and by Guleke, both of which are highly characteristic, as well as the female cases of Gowers,³³ and others (see below). Recently a typical case in a woman who subsequently developed disseminated sclerosis has been briefly recorded by Jacobsohn. This gives us 6 cases in the female sex, to set against 30 male, a proportion of exactly 5 to 1 in favour of the latter.

There fall also to be included in this same clinical group those apparently consecutive to an attack of epidemic encephalitis, viz. the cases of Stiefler,⁸⁶ Missriegler (this case, of much interest, has been seemingly overlooked by both German and English workers), Adie, Münzer, Symonds, Stiefler⁸⁷ (his Case 2), and my fourth case—seven in all, and all males. This raises the sex ratio to over 6 to 1.

In every one of these 43 male and female cases (I do not doubt that I may have overlooked some others unintentionally), the clinical record specifically states that in addition to recurrent diurnal attacks of paroxysmal sleep the individual concerned exhibited in greater or less degree the phenomenon of loss of tone in the skeletal muscles (in one or other part), under the influence of stimuli of the emotional series.

II. Taking next the second subdivision of the first class (and here I shall include both those cases in which the "toneless" attack has been looked for but not found, and those in which no reference is made to it), we find numerous instances in both old and recent literature. The recent cases are those of Adie, Redlich⁶⁹ (his Case 3), Goldflam (2), Curschmann and Prange (their Case 1), Janzen (his Cases 1 and 3); and, among the older ones, after having read through all the original records, I shall include those reported by Chavigny, Foot, Morton, Caton, Ewen, Pitres and Brandeis, and Rybakoff—all in males. The female cases of this subdivision comprise those of Gowers,³³ Blodgett, MacCormac, and possibly that of F. Fischer. In all of these, male and female, no cause for the narcoleptic manifestations was discoverable. But when we come to similar cases in which an association with a definite pathological state seems established, the numbers increase materially. Of the postencephalitic cases we may enumerate those of Stiefler⁸⁷ (his Case 1), Perrier (2), Wenderowic (his Case 2), and Redlich (this

case appears to me uncertain) ; typical cases developing on a basis of commotio cerebialis have been published by Lhermitte ⁴⁶ ; others as clearly linked to one or other variety of epilepsy are those of Féré, Gowers,³² Worster-Drought, Goldflam (his Case 5), Brailovsky (some others here are omitted as being rather indefinite) ; cases arising on a hysterical or other psychopathological basis have been reported by Camuset, Ballet, Laehr, McCarthy, Mammack, Carlill, Myers, Worster-Drought, Brailovsky, and many more ; a syphilitic infection was thought to be responsible in one of Lamacq's cases. It would be easy indeed to enlarge still further this class of recurrent diurnal narcolepsy of comparatively brief duration, for I have made no reference to arteriopathic hypersomnia, or to the sleep attacks of certain forms of drug-taking, or of certain cases of cerebral tumour. Most of the cases of the latter two categories, however, come in reality under our second clinical division. What impresses the student when he surveys this large class is, that as far as recurring attacks of irresistible diurnal sleep are concerned there is no affinity between them and any one specific, invariable form of disease.

Seeing that brief attacks of diurnal sleep can and do occur as a monosymptomatic variety of narcolepsy, it becomes a question of considerable interest whether the *Tonusverlust* that develops on the heels of strong emotion may not also be found monosymptomatically ; if so, what place is it to be assigned in the narcoleptic category ? As far as I can ascertain from a complete survey of the literature, no case of attacks of loss of power or tone through the influence of emotion, occurring by themselves, is on record.* But attention may be directed to the following clinical data.

(1) In Somer's second case, that of a youth of 20, the "toneless" seizures took place not only under the influence of emotion but also after the patient had been walking or standing for a while, being then unaccompanied by any obvious affective experience ; further, they were noticeably more prominent in the picture than the attacks of sleep, which by comparison were few and far between.

(2) In Symonds' postencephalitic case the two types of attack occurred at different periods, for when the patient came under observation he was suffering only from the "affective" seizures,

* Since this paper was written a case of cataplexy has been published by R. D. Gillespie (*Jour. of Neurol. and Psychopath.*, July, 1927) in which "affective attacks" occurred practically alone. Yet sleep apparently of abnormal quality was an occasional afternoon occurrence.

the sleep seizures having ceased and being replaced by insomnia. Conversely, the illness in Hilpert's case commenced with the development of affective attacks, which eventually disappeared although the sleep attacks continued.

(3) Berliner, Hoff and Schilder have recently described a case of monosymptomatic *Tonusverlust*, which they reckon "in die nächste Nahe zum Tonusverlust des Einschlafens und damit zum Tonusverlust der Narkolepsie." The attacks in this case (an organic one) appear to present an intimate similarity to those of the narcolepsy group, but sleep attacks did not occur.

Clearly the effort to circumscribe the conception of narcolepsy alluded to above becomes more dubious than ever if cataplectic seizures can appear by themselves, or be a feature of other states than narcolepsy proper. Further, in this same connexion, the importance of *larval or incomplete varieties of narcolepsy* must be underlined. The following personal case, recently observed, seems to me very instructive.

CASE 5.—*Male case ; larval or incomplete variety. Duration, three years. Cause unknown.*

H. G. B., age 32. Occupation, business.

History of present illness.—For about three years the patient has noticed the occurrence of peculiar attacks, of brief duration, during which he seems to be unable to move his eyes ; he finds himself staring at some object, and in a kind of day-dream ; the object then becomes indistinct, as if seen through a haze, and only by a strenuous effort can he cut short this state and break into its fixity. If no effort is made the phenomenon disappears spontaneously within a minute or two. He has also observed that on occasion this immobility seems to spread so as to involve other musculatures, giving him the feeling that he is unable to move any part of his body, yet at the same time the thought comes into his mind, "Oh yes, I can move if I like" ; as a fact, however, he always remains motionless for an appreciable period, measurable in seconds. No emotion of any conscious kind initiates these spells.

For about the same time (three years) he has repeatedly noticed that under the influence of any excitement or other emotional stimulus he turns deathly pale and loses control over his limbs ; in his own graphic language, "I feel as if I had no knees" ; when playing a game at bridge, wondering what sort of a hand he is going to pick up, he feels his legs "disappearing" and is thankful he is sitting on a chair. With some five or six friends he meets regularly at a restaurant for coffee, and the decision as to who pays is reached by one of the circle piercing the menu card with a pin haphazardly ; the letter nearest the pinhole is taken, and the letter of the respective surnames of the members nearest that in alphabetical order decides. During this trifling procedure he has frequently turned "as pale as a sheet" and, standing on one occasion, had to hold on to the furniture as his lower limbs were sink-

ing. His friends have noticed his demeanour and uncharitably attributed it to unwillingness to pay, whereas in fact it has been the outcome of the affective experience. Hearty laughter produces the same result.

On no occasion, as far as can be ascertained, has the patient had sleep attacks; yet many times during the day he suddenly becomes conscious of "a warm feeling round his eyes," leading him to close them for a moment, and he remarked spontaneously that he always feels that if he chose to he could then allow himself to sleep. As a fact, he has never thus succumbed to the impulse.

Family and previous history.—Negative.

Condition on examination.—A normal finding in all respects.

Had our knowledge of narcoleptic syndromes not widened in recent years I doubt whether the real nature of this case would not have escaped me; as it is, I confidently assign it to an abortive or incomplete class in the general group, at the same time stressing the point that *only the affective or "Tonusverlust" attacks have passed into a characteristic stage.* And it is a further matter of no little interest that this patient also exhibits almost a cataleptic immobility (cf. my Case 3 above) which, brief or momentary as it is, is apparently independent of emotional excitation.

III. If we take next sleep seizures of a prolonged or continuous kind, we are familiar with their occurrence under equally variable pathological circumstances. Clinical distinction here between natural sleep and states of stupor (anergic, catatonic) or of semi-coma is often enough difficult to establish, and the same is true of cataleptic or trance-like conditions and of those associated with drug addiction. Prolonged hysterical sleep is well known and has been the subject of numerous communications (e.g. Raecke); in this connexion no recent writer has mentioned the case of the "sleeper of Oknö," as narrated by Fröderström, who slept for thirty-two years. The curious will find in the volume by Gould and Pyle valuable historical references to cases of protracted sleep of the hysterical, cataleptic, and stuporous kind. Similar more or less continuous, yet sometimes periodic, sleep states also accompany some cases of such organic intracranial conditions as cerebral tumour or abscess; they have been known for many years, and have been made the subject of numerous useful studies (Righetti, Franceschi, Soca, Purves Stewart; more recent communications are those of Bychowsky, Pette, Lucksch; Thomas, Jumentié and Chausseblanche; Souques, Baruk and Bertrand).

Finally, I need here only mention the well-known short or, much more commonly, protracted sleeping states of epidemic encephalitis

and of trypanosomiasis to complete the catalogue of the narcoleptic syndromes belonging to the second clinical group.

Clinical Symptoms

(1) SLEEP

Although the term "sleep" is freely applied to the phenomena of the great majority of the cases usually classed as of narcoleptic character, the ostensible reasons for this require to be examined. It must be apparent to any critical mind that no definite clinical distinction can be drawn between degrees of sleep, which range from merely "dosing" with the eyes shut, on the one hand, to a profundity of unconsciousness not easily to be separated from a comatose state, on the other. In clinical practice distinctions and contrasts are far less easy to draw than they are on paper. Can sleep as a fact be readily differentiated from sleep-like states, and if not, where are the latter to be considered as beginning or ending? In the direction of deepening unconsciousness one passes by easy gradations from somnolence to sopor, sopor to stupor, and stupor to coma, as Monakow long ago emphasized; but if the sleep of narcolepsy is thought to be distinguished by the fact that the patient is easily roused therefrom this is in the first place *not* true for every case, and in the second characterizes some other unconscious states not of the same nature, e.g. some forms of hysterical fit. If, again, we take the descriptions of cases assigned to the group of narcolepsy it is at once evident that in quite a number of instances the attack of sleep does *not* correspond to ordinary conceptions of that state, for we note that frequently the patient is declared to "have heard what was being said," or "to have been aware of what was going on round about him" (Goldflam, Westphal, Redlich, Somer, Chavigny, Mendel, and others). Even a normally light sleeper is not in point of fact asleep if he is cognizant of what is going on. It thus becomes more than doubtful whether the sleep of the narcoleptic is to be likened in all respects to normal, healthy sleep.

Equally remarkable, and somewhat disturbing to any view of narcolepsy as just an abnormality of ordinary sleep, is the significant observation made by not a few that the patients declare they exhibit only the appearance of sleep; they are in reality aware of their surroundings, *but not able to move a single muscle or articulate a single word* (Redlich,⁶⁹ Guleke, Bruno Fischer, Loewenfeld, and others). That is to say, the condition bears only an apparent

resemblance to sleep and turns out to be transitional to the cataleptic class of seizure. In my Case 2 the attack of tonelessness looked rather like sleep, inasmuch as the patient's eyes were closed and his head nodded on his breast, whereas as a fact he was conscious and awake, and the physiological phenomena were of a peculiar kind. Again, in my Cases 3 and 5 trance-like states occurred, apart from emotional excitation, which bore the external marks of sleep, but their content was different, seeing that in point of fact the patient was conscious of his surroundings but unable to move a muscle. The "sleeping" and the "toneless" attacks are evidently linked to each other, in many cases, more intimately than appears from a casual survey. Thus this interesting condition, found in many cases of narcolepsy, widens the conception and approximates it to that of cataleptic and trance-like states, to which some erroneously think it is altogether foreign. This aspect of the problem is of considerable significance.

Another argument can reasonably be advanced against the hypothesis that the sleep of narcolepsy is ordinary sleep, pure and simple. The normal subject falls asleep because he is sleepy, whereas the great majority of narcoleptics are not conscious of undue sleepiness as a prelude to the attack of sleep; nor, for that matter, curtailed as the duration of the sleep nearly always is, are they sleepy or drowsy thereafter; on the contrary, most of them say they are at once quite fresh. And again, the vast majority enjoy sleep of normal duration at night, and never oversleep themselves. It is obvious, in short, that the abnormal slumbers of the day differ from nocturnal sleep in several respects, the possible significance of which it would be well not to underestimate. Suddenness of onset of the diurnal sleep has characterized very many cases; two of my patients have repeatedly fallen asleep at meals, with their food half-way to their mouth. A forgotten case reported more than seventy years ago by Graves is that of a gentleman who had sudden attacks of unconquerable sleep; at meals he broke one wine-glass after another by falling asleep after the act of filling the glass and during the time he was raising it to his mouth—a literal slip 'twixt the cup and the lip. This abruptness of onset is highly characteristic. There are but few exceptions to the generalizations just mentioned. Persistent sleepiness is distinctly rare, as is physical tiredness. Caton's patient had a constant sleepy look and could only keep himself awake by active

exercise or mental stimuli. Tiredness preceded the attacks of sleep in the cases of K. Mendel, Goldflam, Jolly, and some others. In a few cases the conditions and circumstances tending to lead to sleepiness in normal persons (monotony, warmth, post-prandial repletion) have been noticed to affect narcoleptics also, but in numerous other instances no such association has been found.

Because of these peculiarities and differences we are compelled to question any strict identification of the phenomena of narcolepsy with those of normal sleep in the healthy individual, close though the superficial analogy may be, and frequently is. Had we any objective criteria to fall back on, differentiation might be more readily accomplished, but the sign of contracted pupils in sleep, relied on to some extent by Goldflam because noted by him in his cases of narcolepsy, has not a universal applicability. If the useful researches of Richter on changes in the electrical resistance of the body during sleep are corroborated, it may be that along these lines light will be thrown on differences between normal sleep and certain pathological conditions nearly resembling it.

Enough has been said, however, to cast doubt on all simplistic interpretations of the diverse sleep phenomena of narcolepsy—phenomena not embodied in the terminology—and to suggest we are dealing with syndromes not consisting in recurrences of natural sleep but presenting features which offer a link of union with other psychopathological conditions of particular interest.

(2) ATTACKS OF LOSS OF TONE AND POWER

This feature of our first variety of narcolepsy is of consummate interest, because, or in spite, of the fact that it bristles with difficulties.

In the first place, subjective descriptions apart, clinical observation reveals modalities and divergencies of considerable importance. Perhaps the commonest type of attack is that in which, under the influence of emotional excitement, a toneless and powerless state of the skeletal musculature supervenes, with the consequence that the person concerned finds his knees giving way and he sinks to the ground; after a brief period, measurable usually in seconds, tone and power return and he is able to get up again. Attacks corresponding to this general account have been remarked in all of the 43 male and female cases which I have collected, but they vary materially in degree and extent. Sometimes the patient is conscious of no more than a sinking at the knees, possibly scarcely noticeable

to the onlooker ; in other instances the seizure is complete, affecting neck, trunk, arms and legs, and lasting for several minutes. In one and the same case the degree may differ at different times and under differing circumstances.

The actual clinical state is one of transient diminution or loss of volitional power and of muscle tonus, and it is for practical purposes difficult to separate the two components. Objective examination reveals both a loss of power, amounting to absolute immobility, and a loss of tone ; the unique observation made by myself and described fully on a preceding page is of high significance, for it furnishes proof of tonelessness so profound as to result in temporary abolition of the deep reflexes. Numerous observations point to the occurrence of partial and segmental failure of power and tone. The incidents mentioned in my Cases 2, 3, and 4 are paralleled by analogous occurrences in those of Janzen (his Case 2), Wenderowic, Goldflam, and others. In all of these power went out of the limb or limbs about to be innervated volitionally—a kind of motor inhibition under the influence of excitement.

A second feature of the cataplectic attack is that in various recorded cases something more than, or different from, pure loss of tone takes place. Trembling (Westphal), movement of the head and protrusion of tongue (Noack), twitching movements of eyelids and face (Bruno Fischer), of lower jaw (Wenderowic), and other involuntary movements (Redlich,⁷⁰ Carl Camp), have all been seen. The possibility, also, of *voluntary movements of correction* complicating the muscular weakness is specially mentioned by Adie. The speechlessness of many cases is not encountered in all ; my patient (Case 2) made a grunting noise as his head fell forward ; Somer's patient muttered something unintelligible. An equally interesting observation is that of Redlich,⁶⁹ who in one case noted that hearty laughing led to an attack of powerlessness which culminated in involuntary relaxation of both vesical and rectal sphincters.

One of the integral characteristics of the phenomenon is the full conservation of consciousness during its course, however much in some cases the appearance of the patient may belie this.

The exciting cause of the attacks is declared almost universally to reside in a stimulus of the emotional series, among which anger, annoyance, compassion, anxiety or agitation, amusement or joy may be specially mentioned. The *act* of laughing perhaps provokes the attack more readily than any other cause, but in this case the emotional

stimulus is by itself insufficient; anger, and excitement generally, will precipitate the attack, but a stimulus arousing laughter will not produce it should the patient be able to inhibit the *motor act* of laughing. A consideration of the clinical data undoubtedly bears this contention out. In a number of cases it is expressly stated that only irresistible laughter culminates in the loss of power. Only when Gélineau's patient "riait aux éclats" did his limbs give way. My patient, E. C. (Case 2), smiles readily enough, and laughs softly, without experiencing the cataplectic phenomenon. Stiefler's ⁸⁵ patient never developed the affective attack from laughing, but only with rage or annoyance.

Somatic accompaniments indicative of the emotional disturbance, whatever its modality, are not referred to in the literature to any extent—a curious point worth passing notice. I can find only the briefest allusion to these. One of Redlich's ⁶⁹ patients said that in the seizure he had a feeling of "losing his breath"; another said he was conscious of a "warmth spreading over him." In Wenderowic's case the patient was aware of a "pulsating noise in his head"; Somer's second patient showed pallor of the face, as does one of mine (Case 5). The case recorded by E. Mendel concerned a patient who exhibited attacks of *Tonusverlust*, during which there was pallor, the pulse was slow, and perspiration broke out at its close. My own patient, T. T., described with great clearness palpitation of the heart and a feeling of tingling over the head, cheeks and body. Doubtless somatic phenomena of these and similar kinds occur more frequently than is recorded, yet the speculation arises whether, in the case of cataplexy, the emotion does not manifest itself abnormally, to the relative exclusion of the usual outward accompaniments.

(3) TRANSITIONS AND SUBSTITUTIONS

Cursory consideration of the two types of attack might yield the impression that they consist of forms essentially dissimilar, correlated only through their occurrence in one and the same case. This is without doubt an erroneous conception, as can at once be realized by attention to the following clinical variations.

*Emotional excitement may lead not to the cataplectic but to the narcoleptic state.** This occurred with regularity in the cases reported by Kahler and Westphal. Under the influence of emotion the patient whose case is reported by Bruno Fischer felt a strong

* In a case reported by Féré, mentioned on p. 265, uncontrollable laughter was followed by an irresistible desire to sleep.

inclination to sleep. The connexion is shown best of all in Gélinau's original case : " s'il a une émotion profonde, pénible ou joyeuse, le besoin de dormir est encore plus impérieux et soudain."

Sleep may be associated with or follow the cataplectic state. During the attack of powerlessness following laughter, Matzdorff's patient felt an irresistible desire to sleep.

If sleep is prevented the cataplectic attack may ensue. This interrelation is exhibited by my patient E. C. (Case 2 above). Similarly, in the case of T. T. (Case 3 above), interference with the impulse to sleep led to the oncoming of the " paralysing attack," to use the patient's own expression.

The cataplectic state may develop spontaneously, as does the narcoleptic attack in most cases, and be unaccompanied by emotion. This occurred in the cases of Goldflam and of Somer (his Case 2), and in my Case 3, as we have seen, the " paralysing attack " bore no relation to emotion, as far as could be ascertained. A similar remark is applicable in respect of my Case 5.

Many, both of the sleep and the cataplectic attacks, are in reality identical in comprising a component which corresponds to the usual conception of catalepsy or trance. In a previous section I have furnished evidence fully bearing out this statement ; awareness of surroundings, coupled with inability to move, has marked one or other type of attack in the cases of Nammack, Westphal, Somer, Guleke, Gélinau, K. Mendel, Goldflam, Redlich,⁶⁹ Chavigny, Bruno Fischer, Loewenfeld, three of my five personal cases and others. The association occurs with impressive frequency, and cannot be dismissed as accidental.

Consideration of these various divergencies must in my opinion compel us to envisage the possibility that a common factor may yet be found for all the clinical phenomena ; that some not as yet clearly understood inhibitory mechanism is responsible for both the " sleeping " and the " falling " ; that the relations between narcolepsy and other clinical syndromes now to be examined are so close as to render strict delimitation of the former both impracticable and inadvisable.

Relation of Narcolepsy to other Clinical Syndromes

(1) **Relation to Myoplegia.**—As far as I have discovered, Wenderowic is the sole author who makes reference to the resemblance between the attacks of *Tonusverlust* and those of myoplegia paroxysmalis or periodic family paralysis. Yet the analogies are impressive,

in spite of considerable differences. The cataplectic phenomena of my Case 2 at once suggested a certain similarity to those of myoplegia, since in their essentials they consisted of a sudden, widespread, flaccid "paralysis" of skeletal muscles with complete loss of the deep reflexes. The time was much too short for any investigation of electrical reactions; some future observer may be able to furnish us with data in this respect. In the usual family myoplegia the attack is *crescendo* in character, taking, perhaps, hours to develop and hours to die away, but at its height the combination of flaccid palsy with loss of deep reflexes is identical with that reached in a few seconds in the case of cataplexy. I purposely do not emphasize these resemblances, the divergences being obvious to every one, yet it is well to make a note of them; according to Wenderowic a relative lymphocytosis characterizes myoplegia, and a similar blood-picture has been found in narcolepsy by Janzen and by Stiefler.⁸⁷ The significance of this blood-change is as yet indeterminate.

(2) **Relation to Catalepsy.**—This term, like too many other medical expressions, suggests a seeming precision which, as a fact, it does not possess. By catalepsy is usually understood a state of motionlessness, of sustained immobility, with or without clouding of the sensorium; the patient exhibits the appearance of sleep, is commonly indifferent to stimuli, and often shows the phenomenon of *flexibilitas cerea* in the limbs. As a symptomatic term the word has its value, indefinite though it be; the difficulty is that both catatonia and catalepsy convey the idea of the presence of the *flexibilitas cerea* just mentioned; yet this is not a cardinal, in the sense of exclusive, feature of the condition, for I have seen it in epidemic encephalitis, in frontal lesions of organic character, and in other intracranial states (cf. Brissaud⁶). We need an expression to signify the condition of being "struck all of a heap," of immobility following on affective shock, which is sometimes met by use of the phrase "*catatonia attonita*" (cf. *Schreckkatalepsie*). Doubtless catalepsy covers a multitude of dissimilar syndromes, but its main connotation, as given above, has received general acceptance. Many catalepsies are hysterical; the majority, indeed, are, widely speaking, psychopathological. Now we have already seen how many of the attacks of the narcoleptic are signalized precisely by this trance-like state of awareness of environment coupled with inability to move a finger. No allusion is made by any modern writer on narcolepsy to the once much-discussed question of

nocturnal paresis or *paralysis*, which formed the subject of an interesting study by Weir Mitchell.⁵⁶ While doubtless here also a number of differing clinical conditions are comprised, among them is one to which I have long given attention. I have found it in various persons, healthy and otherwise, and do not regard it as anything else than a transient physiological disorder. It consists, briefly, in a state which supervenes commonly in the morning; the subject is awake, because he is conscious and knows where he is, but for the moment he is incapable of innervating a single muscle. He lies in appearance still asleep, with eyes closed, yet within is engaged in an intense struggle for movement, with which is associated an acute mental anxiety; could he but manage to move a limb through a fraction of an inch, the spell would be instantaneously broken, and with a sigh of relief he would regain full power. I have had graphic descriptions of this "sleep paralysis" from more than one patient.

Its resemblance to the "paralysing attacks" of my third narcoleptic (T. T.) is of the closest. It is a physiological catalepsy, a state of sustained immobility with sensorial awareness, and as such is of much interest for our subject. In a large number of the recorded cases the remark is made that the patient can be roused from his narcolepsy or his trance-like condition by a touch, a shake, a movement, whereas stimuli through other sense avenues are less effective. It seems to me this observation may have possible significance.

(3) **Relation to Epilepsy.**—Much controversy has raged round the question of the relation, if any, of narcolepsy to epilepsy. On the whole, most authors deny any connexion between the two. Curschmann and Prange go so far as to advocate abandonment of the term narcolepsy, because "it suggests a relationship to epilepsy which does not exist." Adie repeatedly stresses the point, declaring that it is highly important "we should distinguish it [narcolepsy] from a serious disease [epilepsy] with which it has no connexion whatever." A similar attitude is assumed by Kahler, Goldflam, and a number more.

But there is much to be said against differential absolutism of this kind.

In Chapter I (to which the reader is referred) I have advanced considerations, developed along lines suggested by Hughlings Jackson many years ago, which indicate the possibility of any motor cell-group becoming the seat of a "discharging lesion," so that an epileptic attack may take a number of seemingly very different

clinical guises. The diseased condition (whatever it be) is the cause of the "epilepsy," which is a symptom pure and simple, and, as such, strictly physiological. Adopting this view of the question, I maintain strongly that *any alleged contrast between the narcolepsies and the epilepsies is merely a contrast between symptoms and leaves untouched the problem of whether or not there is a similar mechanism of production behind each.* That there may be, in point of fact, some such similarity of inhibition or release of function is surely conceivable and therefore worthy of discussion. This pathogenic aspect is dealt with later.

The clinical observer must admit general resemblances between the two classes of symptom, which, as Hilpert has recently remarked, "sind in jeder Hinsicht offensichtlich." It is indeed impossible to overlook the periodical recurrence of attack, abrupt onset with or without aura, loss or diminution of consciousness, relatively brief duration; like many epileptics, many narcoleptics are fresh again soon after. The "fit" of sleep is in these respects like many "fits" of another kind. Of course the actual *content* of the "fit" is different in the two cases, but this does not affect the argument; for that matter, as I have said elsewhere (see page 3), the word fit "might be legitimately employed to designate certain attacks of which motionlessness is the prominent feature." Besides, the involuntary movements of some at least of the attacks belonging to the cataplectic series offer more than just a casual resemblance to those of petit mal or of some mild epileptic seizure. Symonds observed one of these in his case, and took it to be epileptic, like petit mal; and I have already mentioned at the outset of the attack in my Case 2 the noise in the throat, the nodding forward of the head, the sinking down in a seemingly unconscious state—phenomena the analogy of which with some of those usually called epileptic it is pointless to try to controvert. If behind the ordinarily great dissimilarity of the external manifestations there is reason to postulate the action of not dissimilar mechanisms, it becomes unscientific to argue for fundamental differences between them from a nosological standpoint.

Amplifying this last consideration, we may examine one or two of the arguments advanced by the supporters of the "no relation" theory. Adie says, "a most important feature of idiopathic epilepsy is that the attacks come on without apparent cause, whereas in narcolepsy the cataplectic attacks are 'reactive,' i.e. there is a definite exciting cause, namely, emotion." And again: "In genuine epilepsy the attacks are never reactive; this feature alone

is sufficient to show that the two conditions are fundamentally distinct." But we have already noted that cataplectic attacks may develop spontaneously (Goldflam, Somer), while the trance-like states of my Cases 3 and 5 also thus developed without any emotional antecedent. In the second place, the view that idiopathic epilepsy is never reactive is contradicted by familiar experience. We need not go so far as Rows and Bond, who explicitly affirm that "a survey of a large number of cases (of epilepsy) . . . has demonstrated that some emotional state involving a disturbance of consciousness, and some reaction to express the emotion, have been found in every instance." But of the occurrence of what we may call reactive or psychogenic epilepsy ("affect epilepsy") most observers have had abundant proof (see page 32); it is recognized by all modern writers on the subject (notably Pierce Clark), and a good illustrative example was recently published in a paper of my own.⁹⁴ This particular criterion of differentiation, therefore, does not serve the purpose.

More important, as forming a positive argument, is the fact that epilepsy and narcolepsy are on occasion definitely associated. For example, one of Gowers' ³² cases was that of a girl, age 14, with a family history of epilepsy, who had suffered for four months from attacks clearly of that nature. But she had also, during the same time, "attacks of sudden pure sleep." When bromide was given regularly the epileptic seizures ceased, but the attacks of sleep went on unchanged. Failure of anti-epileptic remedies in narcoleptic cases cannot, of course, be advanced as an argument for the essential dissimilarity of the two conditions; some epileptics do not react to anti-epileptic drugs. Since the phenomena of narcolepsy are largely akinetic we should not expect sedatives to be the appropriate treatment. A valuable case is recorded by Worster-Drought in which the patient, a man of 25, suffered from fits in childhood up to the age of 10; since 1913 three fits ("description suggesting true epilepsy") have occurred, and also numerous narcoleptic attacks of variable duration, characterized by his falling asleep "at any time, under all conditions, and often in perilous situations." One of Goldflam's patients (his Case 5) suffered from narcolepsy for six years, and later developed typical epilepsy with convulsions. Other instances of the same association are specified by Brailovsky from records not accessible to me; the older cases of Féré must also be alluded to in this connexion, especially his Case 3, that of a youth of 21, who had both epileptic and narcoleptic seizures, the latter

closely resembling normal sleep of brief duration (a quarter of an hour or less). Féré specially noted that as the epileptic attacks increased the others diminished in frequency, and vice versa. In the chapter on "Epileptic Variants" (page 28) is recorded a personal case exemplifying a transition from epileptic to narcoleptic attacks.

During an ordinary major epileptic attack the subject is unconscious, while motor mechanisms are stimulated into abnormal activity; during many narcoleptic attacks the subject is conscious, while the activity of lower motor mechanisms is in the completest abeyance. There is here, as I consider, a certain approach to the existence of "corresponding opposites," in Jacksonian phraseology, which may yet prove to be pregnant with meaning for an understanding of their physiological counterparts. Again, during the examination of E. C.'s (Case 2) affective attack I was able to demonstrate the temporary occurrence of an extensor response. This is known to characterize similarly many epileptic seizures, and it has, further, been observed in normal subjects during sleep. Collier found in a number of healthy children up to the age of 12, during deep sleep, a typical "infantile response" (that is, extension); directly the child awoke the normal adult plantar reflex (flexion) occurred on stimulation. It cannot be without significance for our problem that a "physiological shift," as I call it, of the same type as is seen in normal sleep and in the epileptic fit, characterizes also the cataplectic seizure of the narcoleptic.

In face of these clinical observations it would be a mistake alike to ignore similarities and stress differences; the correct attitude in my opinion is to suspend judgment and to await further contributions to our knowledge. So far, however, as the preceding considerations take us, they make out a *prima facie* case for the possibility of some of the narcolepsies and of the epilepsies having a common origin.

(4) **Relation to Oppenheim's Lachschlag.**—Under the term *Lachschlag* Oppenheim described two cases (one in a girl and the other in a man, age 45) of falling to the ground under the influence of hearty laughter. From the description furnished by the relatives (for no attack was personally observed) he supposed the patient was unconscious during the few seconds only of which its duration consisted, though he says in reference to the first case, "ob volle Bewusstlosigkeit dabei eintritt, haben sie [the relatives] nicht

festgestellt." Adie is inclined to look upon this *Lachschlag* as distinct from narcolepsy, whereas Jolly and Matzdorff both are impressed with the analogy it offers to the *Affektanfälle* of the latter condition; Redlich⁶⁸ points out that the loss of consciousness in Oppenheim's cases was not absolutely certain, and mentions the cases in which sleep follows emotional excitation (see above); Goldflam, too, considers the resemblances of some significance.

I have myself observed one good case of the condition, that of a man of 42, whose symptoms supervened on an injury to the head in the right frontal region. He noticed that when he laughed too loud he tumbled down, a condition which gradually so established itself that he had "dozens of tumbles." If he laughed slightly, he felt a sort of staggering feeling in the limbs; the louder he laughed the more he staggered; eventually he would fall to the ground, but would be on his feet again in a few seconds. Occasionally he "would go right off in the attacks," by which expression I understood from his description that he became unconscious of his surroundings for a moment or two, but not having seen the patient in such a condition I am unable to determine the exact state. By way of defence, he said to me, "I have cultivated a giggle to avoid the laugh." He had no attacks suggestive of sleep, and objective examination was negative.

The analogies which *Lachschlag* presents to the cataplectic attacks of the narcoleptic are much more impressive than the differences, so much so that it may fairly be considered a larval form of the other, or, indeed, more or less identical.

Etiology

The search for possible causes of narcoleptic syndromes has revealed in some instances preceding factors to which the symptoms may legitimately be attributed, but in others none of recognizable pathogenic quality has been discovered.

As regards the former, *head injury* was noted in cases reported by Kahler, Redlich,⁶⁹ and Singer, and occurred also in my Case 1 above. The appearance of narcoleptic phenomena after *commotio cerebialis* is mentioned by Souques, and has been made the subject of a careful study by Lhermitte.⁴⁶ The symptoms in the personal case of *Lachschlag* already mentioned followed an injury to the frontal region. *Alcoholism* in patient or parentage was a feature in those of Noack, Redlich,⁶⁸ Stiefler,⁸⁵ and Hilpert. The possibility of an *endocrine* etiology is suggested mainly by the interesting fact that

in a considerable number of instances narcolepsy has been associated with a definite *increase in weight*, the result of the patient's becoming more fat (Hirsch, Ballet, Münzer, Caton, Jolly, Stiefler,⁸⁵ Lamacq, Redlich,⁶⁹ Hilpert, Janzen, Curschmann and Prange, and my Case 2). The sella turcica was stated to be on the small side in those of Kahler, Redlich, Jolly, Singer and Janzen. In certain female cases the symptoms were aggravated during the catamenial flow (Fischer, Kahler). A somewhat doubtful case reported by Dercum was associated with an acromegaloid condition. We should mention, too, that a relative lymphocytosis of the blood occurred in cases published by Kahler, Stiefler,⁸⁷ Janzen, and Redlich.⁶⁹

Attention must be directed also to the peculiar connexion of some narcoleptic cases with *epistaxis*.

(1) The patient of K. Mendel suffered from almost daily nose-bleeding between the ages of 8 and 22. When this finally ceased narcolepsy supervened.

(2) In a female case (MacNamara) a girl, age 15, had sudden attacks of profound sleep, which disappeared after a severe epistaxis, and did not return.

(3) Hilpert's patient exhibited the typical combination of narcolepsy and cataplexy. An illness (? "cerebral influenza") developed intercurrently, associated with strong epistaxis, and thereafter the cataplectic attacks ceased altogether, though the narcoleptic continued.

(4) I have recently seen in consultation a case of narcolepsy (with cataplexy) in a young man, with high blood pressure and slow pulse, who used formerly to have bouts of epistaxis regularly, about once a month. Since the onset of the narcolepsy the epistaxis has entirely stopped.

These facts are rather striking, even though their significance at present eludes us.

Again, an origin for the narcoleptic syndrome in some disturbance of a *psychopathological* kind was definitely ascertained in cases reported by Carlill, Myers, Worster-Drought, and Missriegler, and was probable in those of B. Fischer, Morton, Stöcker (his Case 4), and Brailovsky.* This aspect of the problem has undoubtedly been somewhat ignored by neurologists preoccupied with organic disease, and it might be developed further with advantage. I do not

* Gillespie says of his case of catalepsy alluded to in the footnote on page 91, "psychological factors were evident enough, but psychotherapy had no lasting effect."

question that the so-called idiopathic variety might be further dismembered were an adequate psychological investigation undertaken more frequently.

The data which have just been examined, together with what has been said on a previous page, apropos of clinical varieties, are sufficient to show how diverse and heterogeneous is the etiology of the narcoleptic syndrome. To assort the causal factors is a matter of no little difficulty ; a provisional arrangement might be made along the following lines :

- (1) Traumatic group.
- (2) Psychopathological group.
- (3) Endocrine group.
- (4) Epileptic group.
- (5) Toxi-infective group (epidemic encephalitis, etc.).
- (6) Circulatory group.
- (7) Pressure group (cerebral tumours, etc.).
- (8) Group in which no *Grundkrankheit* has been discovered.

Doubtless a cross-division may occur here ; again, some etiological components are surmised rather than proved in particular cases. In others the relation of the syndrome to the apparent cause, or to an outstanding feature of the individual case with which it may be linked, remains obscure, e.g. in respect of epistaxis and diplopia. I have already alluded to the former ; the latter was noted in cases recorded by Janzen, Spiller, Hilpert, and myself (Case 1). Whether it is of etiological significance is undetermined ; according to Janzen it is no more than a "prehypnotic" or "hypnagogic" diplopia, occurring in many normal people if they try to resist sleep. This explanation, however, does not appear to apply to my Case 1, in which the double vision was complained of independently of the sleep attacks, yet so far I have failed to discover any objective basis for it.

For the present the student of narcoleptic symptomatology must be content to recognize the diversity of the bases on which it arises in many instances, and to acknowledge his ignorance of them in others.

Pathology

No autopsy on a case of narcolepsy belonging to the first clinical division has up to the present been conducted, so far as I know. Patients do not die of narcolepsy, which, as we have seen, is no disease.

What pathological information is at our disposal is derived from cases belonging to the second division, viz. cases of prolonged or continuous sleep. The light they throw on those of the first group is oblique and indirect, and by itself but fitfully illuminates an obscure subject.

In 1900 a case of prolonged sleep associated with a basal cerebral tumour was published by Soca; the patient was a girl of 18; during seven months "*la malade dormait toujours, toujours de son sommeil paisible, d'aspect parfaitement naturel, toujours se reveillant facilement aux provocations ordinaires.*" The tumour was subsequently found to have penetrated into the third ventricle, and to have destroyed completely corpora mamillaria, tuber cinereum, chiasma and optic nerves.

Righetti's case (1903), that of a boy of 12, was one in which similar prolonged sleep was by far the most prominent symptom: "*fino dell' ammissione il paziente e in uno stato di sonno continuo. La sue fisionomia appare calma, il respiro e regolare come quello di una persona sana che dorma.*" When called in a loud voice he would wake up, sigh, and fall asleep again. In this instance the tumour completely destroyed optic nerves, chiasma, bandelette, and tuber cinereum, while the walls of the third ventricle were distended. Basing his researches to some extent on statistics furnished by Schuster, Righetti collected 775 cases of verified cerebral tumour, of which 115 were noteworthy clinically for the occurrence of pathological sleep. He compiled the following figures, which I have embodied in a Table.

TABLE I

Site of tumour.	Percentage of cases showing abnormal sleep.
Thalamus and third ventricle	34·7
Medullary.	27·9
Hypophysis and vicinity	26·5
Corpora quadrigemina and epiphysis	25·6
Cerebellum	15·8
Central gyri	10
Frontal	6, etc.

From these figures it is at once obvious that tumours situated in relation to the third ventricle (walls, floor, anterior and posterior extremities) together form a far higher percentage of abnormal sleep cases than any other group, and the association must be of significance.

Purves Stewart published four cases of "tumour in the hypophysis cerebri" in 1909, and of these three were characterized by the occurrence of intense drowsiness, sometimes paroxysmal, lasting for hours, days, or weeks at a time. "In Case 1, and less strikingly in Case 4, the drowsiness had curious intermissions, during which the patient woke up for a time and was apparently clear and alert." From the pathological descriptions the reader will find that while the pituitary itself was not invaded in all, the one common site of involvement was precisely the floor of the third ventricle.

Both before, and since, analogous cases have been put on record; recent German writers on the subject confine their quotations largely to the cases of Pette and Lucksch, which are not as a fact by any means so striking as some of those of which a *précis* is here furnished. Weisenburg collected thirty cases of tumours of the third ventricle and found that "a decided tendency to drowsiness or somnolence was specifically noted in sixteen, most of these being situated in the floor.

The correlation of clinical and pathological findings in cases of cerebral tumour must be undertaken circumspectly; the effects of pressure, of œdema, of toxicosis, cannot be ignored, on the one hand, and, on the other, intracranial growths are commonly in the event too large for conclusions of localizing worth to be drawn therefrom. Thus it is that a tumour away from the third ventricle may be associated with somnolence or hypersomnia; in a case recorded by Bychowski it was situated on the under-surface of the left frontal lobe. Every neurologist can from his own experience quote cases of cerebral tumour of this kind; nevertheless the general conclusion holds, that basal tumours behind the chiasma are more likely than others to manifest themselves, *inter alia*, by hypersomnia.

It is equally hazardous to attempt to draw deductions from cases of cerebral abscess or of cerebral arteriopathy, for reasons which must be apparent. The evidence derived from epidemic encephalitis has been marshalled more particularly by Economo, whose general conclusion is to the effect that lesions in the vicinity of the third ventricle (floor, periventricular grey matter, region of the *iter Sylvii*) are responsible for the disorders of sleep that are highly characteristic of the average case of that disease.

Sleep Centres and their Localization

Pathology can contribute comparatively little *specific* evidence, as we have just seen, but there are collateral lines worth scrutinizing.

Of these one that appears to have some importance is the experimental, though the data are rather meagre even if significant, and also now somewhat old.

Perhaps the most valuable work is that of R. Dubois,¹⁸ whose results were published as long ago as 1896, and have been neglected by recent writers on the subject. Dubois experimented on marmots, and proved that "vers la partie antérieure de l'aqueduc de Sylvius et du côté du plancher du troisième ventricule, il existe des centres respiratoires de ralentissement et d'accélération, d'où dépendent également l'hypothermie et le réchauffement, la torpeur et la veille." He regarded the "centre" as a "centre du réveil," which plays "un rôle prépondérant dans le mécanisme du sommeil." This "centre" regulates respiration and circulation and produces natural sleep by a process of "autonarcose carbonique." Reviewing his experiments in a later publication, the same author¹⁹ states that "J'ai mis hors de doute par l'expérimentation sur les marmottes un centre de sommeil, qui est d'ailleurs le même que celui du réveil, et se trouve justement situé dans la région où les auteurs que je viens de citer ont observé des lésions pathologiques coïncidant avec un sommeil persistant et prolongé." To Dubois without question must be assigned priority for some of the evidence now attributed to more recent work on the third ventricle and its grey matter.

Experiments conducted by Cushing and Goetsch on hibernating wood-chucks dealt mainly with the pituitary gland, and led these workers to attribute hibernation to "a seasonal physiological wave of pluriglandular activity." They found histological changes especially in the pars anterior of the hypophysis in hibernating animals, as had Gemelli some years previously in marmots. The latter, however, did not ascribe the somnolence of his animals to changes in the gland, which he did not regard as a hypothetical centre for physiological sleep. Advances in knowledge of the last few years render implication of the pituitary problematical for these sleep states, and seem to invalidate the speculations of those who, like Salmon, attributed hypersomnia to functional insufficiency of that body.

Evidence has gradually collected in support of the general view already mentioned, a view to which a consideration of certain tumour cases, encephalitis cases, and Dubois' experimental work leads, viz. that physiological centres related to the floor of the third ventricle and the region of the iter—centres in the hypothalamus—have somehow a connexion with the process of sleep. Undoubtedly

much of this localization remains purely hypothetical, though it may well be of heuristic value. In the midst of conjectures (for pathological proof is wanting) the researches of Stief should be given proper weight; that observer has found constant, marked changes in the nucleus paraventricularis and in the hypothalamus generally, in cases of general paralysis, senility, cerebral arteriosclerosis, various kinds of psychosis, cachectic states, etc., and while he believes they explain the development of vegetative symptoms, and possibly the onset of the senium, there is no mention of sleep as forming a feature of any of his cases. For the present, therefore, exact pathological knowledge does not take us very far along the road to a solution of the problem of what mechanism or mechanisms are responsible for the symptoms of narcoleptic patients, or of where precisely they are situated.

One of the major difficulties yet to be surmounted is constituted by our ignorance of the physiological processes underlying natural sleep. It is well not to ignore the extreme complexity of the factors, a point which of itself renders highly improbable the existence of a *solitary* "sleep regulation centre" localized with mathematical exactness towards the posterior-inferior end of the third ventricle. Sleep must be regarded as a complex physiological process, which can be influenced both by physicogenic and psychogenic excitations. Its actual phenomena comprise manifestations belonging to different series—respiratory, circulatory, muscular, sensory and psychical. To suppose all these varying phenomena to be somehow controllable by a relatively limited ganglionic centre puts rather a strain on physiological belief, even if some lines of evidence appear to indicate the possibility of a ventricular nucleus playing a definite though undetermined rôle in the *initiation* of the whole somatic process. We do not know whether this "centre" inhibits or releases, or what precisely are the mechanisms it controls. In view of these facts some of the contentions advanced in recent communications will not bear investigation. Hirsch,³⁸ for example, because his case of pathological sleep was found to be associated with an abscess of the left optic thalamus, says that "sowohl der mediale Teil des linken Thalamus opticus wie das in der Nähe der Okulomotoriuskerne gelegene Höhlengrau Schlafzentren im richtigen Sinne sind"! "Localization" of this crude kind requires no comment.

We may, for the moment, consider Pavlov's views (1927), based on his work on dogs in connexion with conditioned reflexes, and see

how far they take us towards an explanation of the phenomena of narcolepsy. According to him, "sleep is nothing but internal inhibition which has become diffused continuously over the entire cortex and has descended also to some of the lower parts of the brain." Thus he considers sleep to be of cortical origin, but in its usual form simultaneous involvement of infracortical centres takes place, as evidenced by "relaxation of the skeletal muscles (closure of the eyes, drooping of the head, sagging limbs, and body limply hanging in the loops of the stand)." If, however, "a complete inhibition [is] confined exclusively to the cortex, without a concurrent descent of the inhibition into the centres regulating equilibrium and maintenance of posture (centres of Magnus and de Kleijn)," then a different form of "sleep" is observed; the animal "preserves an entirely alert posture . . . with wide-open immovable eyes . . . remaining motionless sometimes for minutes and sometimes for hours. On changing the position of an extremity such extremity retains the new position." In other words, "the animal is in a state of catalepsy."

Interesting as these observations are, to what extent do they illuminate the problems of narcolepsy?

In the first place, as we have repeatedly observed, the "sleep" of the narcoleptic certainly in numerous instances does not conform to ordinary sleep, and in such cases there can be no universal cortical inhibition, with descent to centres regulating equilibrium and posture. Various examples of the narcoleptic keeping on his feet, nay, walking along, when "asleep," might be cited. To hold that in such cases internal inhibition is confined to the cortex, subcortical centres remaining in activity, is illegitimate, for as we have just seen this physiological condition results, according to Pavlov, in catalepsy, which is signalized by absence of movement.

Secondly, Pavlov's experiments do not help us to understand the phenomena of cataplexy, unless by analogy we are to allow that internal inhibition might invade the "centres of Magnus and de Kleijn" while the cortex is as a fact functioning normally, in the sense that the patient is conscious and in a state of awareness. If so, there is here no question of "descent" from cortex to mid-brain, and the speculation is physiologically unsupported, as far as Pavlov's experiments go.

On the other hand, I attach importance to his physiological explanation of catalepsy, which condition I have tried to show is linked to narcolepsy. The passing attacks of immobility,

trance-like states, distinguishing more than one of my own cases and noticed also in others, receive here a possible elucidation.

Pathogenesis

At this point it is well to recapitulate from the clinical aspect the salient features of narcoleptic cases, lest any preconception of what is implied in the term "sleep" should mislead us.

(1) In the great majority of instances the *nocturnal* sleep of the narcoleptic is normal. We must, therefore, consider that we are dealing with mechanisms that are disordered only during the day, and not at night. This fact of itself disposes of the contention that in our first clinical division of narcoleptic cases any structural impairment, or any more or less permanent abnormal physiological state of the mechanisms concerned, is in existence.

(2) "Sleep" is a complete misnomer for the attacks in a large number of cases. In many, the patient is not asleep, but motionless and aware of his surroundings.

(3) A whole clinical group is constituted by cases in which attacks both of sleep and of tonelessness occur. Not only are these in reality less separable one from the other than appears on superficial consideration, but emotional excitations sometimes cause one to develop, sometimes the other. Again, the toneless attacks may supervene without obvious affective antecedent, as may the sleep; on occasion, one may be exchanged for the other.

(4) All gradations are met with, from narcolepsy without cataplexy, to cataplexy without narcolepsy.

(5) An outstanding clinical feature of both types of seizures is their relative brevity, their diurnal periodicity, and the rapidity with which the patient recovers from them.

With these considerations in mind, we may in conclusion attempt an approach to the problem of pathogenesis.

I am in accord with previous observers who have commented on the fact that affective hypotonia is seen from time to time in normal people, or who have referred to the embodiment in colloquial terminology of the existence of emotional influence on body musculature. Affective *Tonusverlust* certainly occurs unspecifically in many ways. From this viewpoint, its occurrence as a feature of narcoleptic syndromes is merely an exaggeration of the normal, and forms a difference in degree and frequency but not in kind. Laughing, tickling, fright, fear, may in the normal person lead to a help-

less, toneless, motionless state of skeletal musculature. German writers refer to the *Totstellreflex* of the frightened animal, "frightened out of its life," as we say. Allusion was made above to the occurrence of sphincter relaxation during the cataplexy in one of Redlich's cases; this also can scarcely be judged pathological if we recall certain slang expressions that seem to be common to many races. A patient under the care of Brissaud⁷ had a similar experience.

Evidently, then, the affective or cataplectic attacks are not essentially pathological, and unlike anything seen in normal persons; on the contrary, the mechanism or mechanisms in activity at such times may on occasion be in action also under conditions that cannot be regarded as abnormal. In my opinion *this* is the main distinguishing feature between narcolepsy and ordinary epilepsy, considered as symptoms; the narcoleptic attack (either narcoleptic or cataplectic) reproduces a phenomenon to be seen also from time to time, in minor degree, in persons of normal neural function, whereas the epileptic attack does not. It is essentially abnormal, and not to be likened to any neural phenomenon seen under normal circumstances. To this differentiation I am inclined to attach considerable weight.

Clearly, in respect of the affective seizures, a mechanism of *general motor inhibition* comes into play, in whole or in part; its outward manifestation ranges from mere trembling at the knees to temporary abrogation of the functions of standing, moving, speaking; whether this universal inhibition closes motility from the side of innervation, or of tonus, or both, is, as I have pointed out, difficult to determine and in one sense relatively immaterial. My examination of the physiological state during one of these attacks has shown the outfall of tonus to be so absolute as to cause disappearance of the knee-jerks. Where is this centre of tonus, whose inhibition leads to universal atonia? Is it labyrinthine, cerebellar, mesencephalic, or pre-mesencephalic? The toneless sinking to the ground is distinctly reminiscent of the effect on the boxer of a blow on the point. To quote Sherrington: "The 'knock-out blow,' where the lower jaw conveys concussion to the otocyst, reduces in a moment a vigorous athlete to an unstrung bulk of flesh whose weight alone determines its attitude, if indeed a reactionless mass can be described as possessing attitude at all." Is there loss of deep reflectivity in the moments that follow the knock-out? Unfortunately I can find no observations to aid in determining the question. Certain it is

that consciousness is not by any means always lost, so that the parallel between this state and that of affective cataplexy is sufficiently close physiologically. If merely for the sake of discussion we postulate a labyrinthine origin for the somatic symptoms, and assume continuance of integrity of the "volitional" motor system which nevertheless is incapable of overcoming the absolute atonia of the moment, by what mechanism does emotion affect the labyrinth or its central representation? Is it conceivable that we have here through disease stumbled upon a biologically old automatism, a defence-reaction in which immobility is of advantage to the frightened animal, "struck all of a heap"? If this should seem a feasible speculation, why is the mechanism activated by the act of laughter in so many instances? Hysterical laughter sometimes covers fear, yet the explanation is surely insufficient otherwise.

But the data of partial or incomplete cataplexies, as already described, appear to show as least as definite an *inhibition of volitional innervation* in the limb concerned as of muscular tonus. Under the powerful influence of the emotion of the moment innervation is inhibited and a kind of aboulia results. These facts would seem to indicate an inhibitory action of emotion over *cortical* activities, no less than that over mechanisms at an *infracortical* level in the complete cataplexies.

In the attacks of "sleep," further, it is permissible to see a generalized inhibition of muscular activity (in numerous cases only the appearance of sleep is exhibited, the essential feature consisting of inability to innervate a single muscle).

Thus the problem of pathogenesis, in my opinion, reduces itself to one of *inhibition*. Inhibition of tone, inhibition of innervation—local or general, cortical or infracortical, or both—here surely is the clue to the diverse phenomena of narcolepsy, cataplexy, and probably also of catalepsy, so intimately interrelated in the cases that have occupied our attention. By comparison with the akinesia and atonia, the occasional involuntary movements of some cataplectic attacks (mentioned on page 97) may be considered escape phenomena, and serve in reality to emphasize the almost universal cessation of motor function.

It remains, therefore, if we bear in mind the epileptic variant which I call akinetic or inhibitory epilepsy (page 42), for future study to elucidate the circumstances under which sensory, affective or emotional excitations inhibit instead of arousing motor activities, as well as those in which "sleep" and trance-like states

develop apparently in independence of such affective stimuli—"spontaneously," as we say.

One narcoleptic state may closely resemble another in externals, but be distinguished pathogenically by the source of its excitant element. In a given instance it may be the outcome of excitations of a psychological order, in others it is a sequel to local conditions of the same level, or to impressions extending from lower levels. And there is no reason to doubt that in the psyche itself there may be levels of function, so that the pathogenic possibilities are correspondingly increased.

Only in some such way as this can we find a possible interpretation for the diverse narcoleptic symptoms of the psychoneurotic, the postencephalitic, or the sufferer from cerebral tumour, as well as of the patient whose case is classed as spontaneous or idiopathic.

Conclusions

At various points throughout this discussion, the conclusions to which it appears to lead have been briefly indicated, and further recapitulation is unnecessary. Their tentative nature, however, must be fully admitted.

With unfamiliar conditions like the "narcolepsy" which some persist in calling a "disease," attempts at identification and at nomenclature should come last, not first. The analogies which various narcoleptic syndromes offer to those usually termed epileptic are here examined without prejudice; differing in outward manifestation, they none the less are such as to suggest that a more or less similar physiology lurks behind each. What precisely this is, however, it is impossible at present to determine.

Again, narcoleptic phenomena on analysis turn out to exhibit affinities to those not only of epilepsy but also of catalepsy, while it is also found that "sleep" is a term used with vagueness and indeed with inaccuracy in respect of many of the symptoms classed as narcoleptic. The suggestion is that greater significance (from the viewpoint alike of etiology and of pathogenesis) attaches to the toneless, powerless, motionless awareness of numerous cases (both in "sleep" attacks and in cataplectic attacks), than to actual sleep.

It is useless to force our cases of "narcolepsy" into the category that seems suited to them unless they fit comfortably; the initial task of minute clinical observation and investigation is still far from reaching its end.

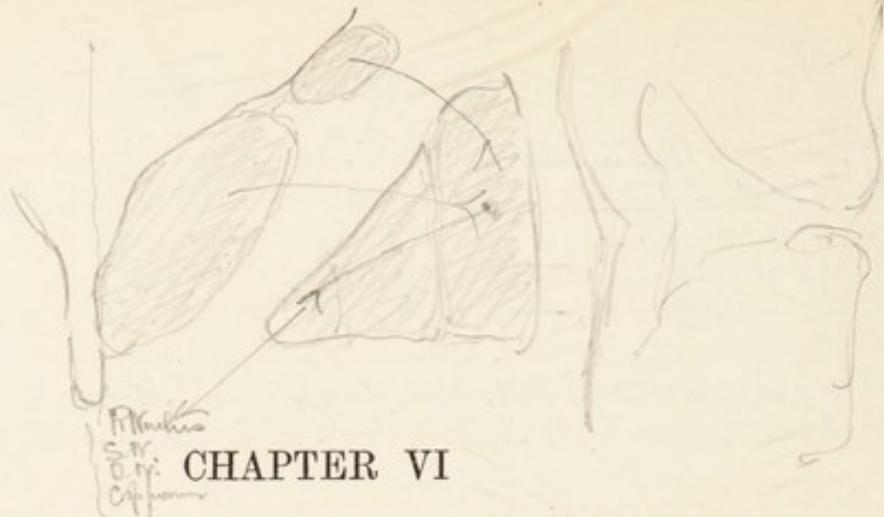
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CHAPTER VI

THE OLD MOTOR SYSTEM AND THE NEW *

Comparative Anatomy and Physiology. Experimental Physiology. Clinico-anatomical Investigation. Disease of the Corticospinal System. Disease of the Old (Striatospinal) System. Simultaneous Involvement of Corticospinal and Striatal Motor Projection Systems.

Among various noteworthy advances made by neurology since the beginning of the century none is more striking than that concerned with our knowledge of motor disease.

A single illustration will serve to demonstrate this progress. In the third volume of Sir William Allchin's *Manual of Medicine*, published not much more than twenty years ago and modern in its outlook, paralysis agitans finds a place under "Functional Diseases," between occupation neuroses and night terrors. To-day, no student of the subject can pick up a journal of neurology, English or foreign, without a likely chance of finding therein contributions dealing with the phasic and static activities of the motor nervous system, with muscle tonus, extrapyramidal motor disease, postures, and involuntary movements. Along with the advance of which innumerable papers are the token, however, there has come increasing recognition of difficulties, or, rather, those who would seek to grasp established truths are still conscious of a good deal of groping. The reasons for this state of affairs are manifold: (1) Many clinical cases are remarkable for the complexity of the motor phenomena encountered. This is particularly the case when disorders of tone and attitude are coupled with both paralysis and involuntary movements. (2) It is difficult often to separate the actions of different mechanisms which may be simultaneously involved. (3) Overschematization of syndromes and rigid ascription of particular symptoms to particular anatomical systems have hindered know-

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ledge, instead of augmenting it. (4) Too simplistic an interpretation of the clinical phenomena of disease by reference to the results of experimental neurophysiology has also been a disadvantage. (5) Experimental results are themselves sometimes in conflict, as are, unfortunately, the pathological findings in not a few cases presenting clinical resemblances or identities.

While the gloom enshrouding many problems of the motor system is in process of being dispelled, the light is perhaps not yet sufficient for recognition of more than the salient features of some of the various mechanisms as they now appear to stand out. In the present communication I shall attempt a brief survey, considering the subject of the old and the new motor systems from a threefold point of view: (1) from that of comparative anatomy and comparative physiology; (2) from that of experimental physiology; (3) from the standpoint of the clinician and the neuropathologist. A succinct account of the present position of knowledge as it appears to me will give an opportunity of indicating in what respects caution and criticism are still clearly needed, and of outlining several personal views that seem to commend themselves.

Comparative Anatomy and Physiology

As an essential preliminary to the study of the clinical phenomena of motor disease we must consider the old and the new motor systems from the standpoint of comparative anatomy and physiology.

In the vertebrate series the familiar corticospinal (pyramidal) efferent motor system only appears when the pallium or cortical mantle develops above or anterior to the original motor centres, which for the moment may be alluded to as "lower motor centres." This development of what later becomes the corticospinal tract, uniting an incipient pallium to the spinal cord, is first encountered in the higher reptilia. In birds such descending fibres are so few as to be negligible, and their pallium (largely a visual and olfactory pallium) has been shown to be electrically inexcitable; hence Ariëns Kappers' ¹ view that in these vertebrates the old motor centres act vicariously for the new. Be this as it may, only when we reach the lower mammalia do a pallial motor ganglion and a projection system therefrom to the spinal cord become permanent anatomical features of the nervous system.

What are the original motor centres of prepallial days? By the term "subcortical or lower motor centres" are signified, loosely,

ganglionic groups at lower levels which, from the histological character of their nerve-cells and from their possessing fibre systems of descending type, are presumed to be efferent or motor in function. The fundamental motor unit in this respect is the lower motor neurone, from ventral horn nerve-cell to end-organ in striated skeletal muscle. Regarded from the viewpoint of phylogeny, it is the original, and simplest, unit of motor activity. The oldest motor centres are the spinal centres. (For our present purpose we need not descend lower in the scale of living creatures than to those in which a central neuraxis can be recognized.) Further, in the whole vertebrate series the basal part of the telencephalon or forebrain forms a prominent organ, known as the corpus striatum and more or less tacitly assumed to be the highest centre of the old motor system, the old "upper motor centre." Evidence goes to show that, for the lower animals (fishes, reptiles, birds), the corpus striatum represents the farthest central motor development hitherto attained. Divisible into a palæostriatum (the globus pallidus of the higher vertebrates), as obtains in fishes, and a neo-striatum (the putamen-caudate of the higher members of the animal series), as found added to the former in reptiles and birds, this fundamentally significant organ may be regarded, in Elliot Smith's ² words, as that part of the original cerebral hemisphere whereby impressions of smell, and other sense impressions, may bring their influence to bear on nervous mechanisms regulating movement. If this is so, it must be a ganglion of a somewhat different order from the central motor cortex. At the outset, indeed, we are faced with the fact that the differences between old and new motor systems, phylogenetically considered, are more striking than the analogies.

(1) We cannot be sure that the functions of the corpus striatum in creatures with little or no motor pallium are comparable to those of the motor cortex of higher animals. On the contrary, we are struck by the fact that there are pronounced histological differences between the two, and that the former, at least in the higher animals, does not, when electrically stimulated, give rise to recognizable skeletal movement. It is conceivable no doubt that when the level of the mammalia is reached the corpus striatum has already changed in function from what it may have in the case of fishes, reptiles and birds. In any case, we do know that it is closely associated, in the most primitive type of forebrain, with olfactory and possibly gustatory impulses, and that part of it is in fact an olfactory cortex

or archipallium. Otherwise expressed, part of it in the history of the animal series must be supposed to exercise receptive or sensory functions. We cannot, therefore, on grounds of comparative anatomy and physiology, consider corpus striatum and motor cortex comparable without a good deal of qualification.

(2) Again, the striatal projection system is not similar to that of the motor area of the cerebral cortex, which is anatomically unbroken to the spinal cord; on the contrary, it is peculiarly complicated. It consists of an original "basal forebrain bundle," a tractus striothalamicus and a tractus striosubthalamicus in one, linking the organ with still lower motor ganglia via the optic thalamus. The original efferent pathway from the palæostriatum is interrupted in the latter ganglion—an important consideration, since clinical attention to the *sensory* thalamic syndrome has led to a certain ignoring of the *motor* disorders connected with disease of that ganglion; moreover, stimulation in apes of the thalamus (particularly of its lateral and the ventral part of its median nuclei) gives progression or locomotion movements—again a significant fact. The palæostriatum, once more, is linked definitely by a tractus striomesencephalicus to the midbrain. Thus the corpus striatum has no direct spinal projection system as has the motor cortex, a datum which of itself suggests that the original functional relation of the old upper motor centres to the spinal cord cannot have been identical with that of the new upper motor centres to the same part of the neuraxis. Indeed, as has just been remarked, movements are obtained experimentally with greater ease and certainty from various points within the optic thalamus than from the corpus striatum. We may legitimately hesitate, therefore, to ascribe motor functions to the latter which can be compared with those even of the former. Striatal efferent paths to the spinal cord are interrupted in optic thalamus, regio subthalamica and mesencephalon. At each of these places are collections of cells from which the links to the spinal cord take origin. The complexity of this projection system as compared with the simplicity of the corticospinal projection system must be kept steadily in view.

Thus the place of the corpus striatum in the old motor system is still rather obscure. The "primitive somatic motor fasciculi" of the lower animals are apparently derived in part from corpus striatum, in part from optic thalamus, or are in contact with efferent systems from both these ganglia, and it is incorrect to imagine the former purely motor in function, the latter purely

sensory. Were there a direct striospinal path to neural mechanisms regulating movement the problem would be easier than it is. In the absence of this, we are perhaps not justified in assigning motor *innervation* to the corpus striatum, though we cannot probably be far wrong in assuming that the primitive somatic motor fasciculi are the homologues of the corticospinal tracts in man, and that these are influenced at their oral origins by impulses derived from the corpus striatum, either directly or via the optic thalamus.

(3) The relation of neostriatum (putamen-caudate) to palæostriatum (globus pallidus) is one of the difficult subsidiary questions in a thorny subject. In animals with poorly developed pallium the connexion of these two striatal divisions with each other is far from clear, and with increase of pallial action the problem certainly does not become easier. By some a parallel has been drawn between the development of the corpus striatum as a whole and that of the pallium; the relation of neostriatum to palæostriatum has been thought analogous to that of motor cortex to spinal motor centres. But the analogy is misleading; we cannot say in actual fact that the function of the palliospinal system usurps that of the striatal system, for, as we have seen, it is doubtful if the corpus striatum is a motor organ of the innervation type; hence we cannot by any motor cortex analogy suppose that the neostriatum abrogates or inhibits the function of the palæostriatum. I mention this now because we shall see later how a speculative theory of differentiation of function as between neostriatum and palæostriatum has been erected on inadequate evidence.

Experimental Physiology

As a second preliminary to our clinical approach we must ascertain whether any experimental distinction can be drawn between the old motor system and the new.

To take the latter first, the phenomena of electrical stimulation of the excitable cortical motor area in man are familiar enough; the musculature of the opposite side of the body is represented from toe to trunk, from trunk to arm and finger, from neck to face, etc., in the precentral gyrus from above downward. We mention, in passing, the well-established facts of reciprocal innervation and of the phasic nature of cortical motor activity. Only so long as the stimulus is applied do the corresponding muscle or muscles contract; with its removal, contraction immediately ceases; there is no after-discharge. This is true, also, of stimulation along the

course of the corticospinal tract. For example, when flexion of the arm is produced by electrical excitation of the appropriate fibres in the pyramidal tract at the level of the crus (in the decerebrate animal) this flexion, as Graham Brown³ says, "climbs" during the course of application of the stimulus. Cessation is followed by a sudden relaxation, so that the arm "flops" in a very characteristic way.

Artificial excitation of the old motor system, on the other hand, leads us at once to problems which are far indeed from being solved.

As regards the corpus striatum itself, investigation of the whole organ with a stimulating needle has failed, in the ape,¹⁵ to elicit any evidence of the localization of motor centres there, comparable to the centres of the excitable motor cortex.* This is true of both palæostriatum and neostriatum, and is of fundamental importance in view of the fact, already mentioned, that movements can be readily obtained from certain parts of the thalamus. It is a legitimate inference that the motor functions of the corpus striatum cannot belong to the order of simple innervation, and we may therefore ask ourselves whether it may conceivably stand to the rest of the old motor system somewhat in the relation of the higher motor cortex (frontal) to the motor cortex proper (rolandic). This point will be referred to subsequently.

It is otherwise with the optic thalamus and the mesencephalon. Allusion has already been made to the progression movements elicited from the median and ventral nuclei of the former ganglion.

In respect of the mesencephalon, noteworthy motor reactions are found in the decerebrate animal. We owe largely to the work of Graham Brown³ our knowledge of this part of the subject. Unipolar stimulation of the cross section of the midbrain obtained by decerebration at the level of the anterior colliculi, at a point dorsal to the corticospinal tract in the crus, constantly produces a definite, specific postural motor reaction on the part of the animal experimented on. The area from which this result is invariably obtained is dorsal in the tegmentum and includes the region of

* Whenever the needle begins to impinge on the adjacent corticospinal tract in the capsule the movements obtained are unmistakable. Occasional claims to have elicited movements from the ganglion itself have been made, one of the most recent being that of Delmas-Marsalet,⁴ who worked on dogs. But his results are open to both technical and interpretative criticism.

the red nucleus, the part of the superior cerebellar peduncle running to it (*tractus cerebellotegmentalis*) and the posterior longitudinal fasciculus. The attitude is as follows: The head is tilted back and also twisted so that the face looks to the side stimulated; the homolateral arm is flexed and the opposite one extended; the leg of the same side, on the contrary, is extended and the opposite one flexed (as a rule); the tail erects and is bent to the stimulated side. The back is usually slightly convex to the opposite side. When stimulation has ceased, the posture may continue unchanged for many seconds, even minutes. From the appropriate area on the opposite side the posture is obtained reversed. The motor activities evoked are essentially postural; they can be obtained by appropriate stimulation after ablation of the cerebellum, but thereafter they gradually lose in duration. They can also be got, and maintained for many seconds, in limbs all the posterior roots of which have been cut months before.

One of the important points for our purpose is to note how, in comparison with these postural activities, that of the corticospinal tract is fundamentally nonpostural, i.e. is phasic rather than static. Experimentation has shown conclusively that pyramidal motor reactions correspond strictly to the duration of stimulus, while extrapyramidal motor reactions do not.

Apart, however, from actual stimulation of mesencephalic foci, the phenomena of experimental decerebrate rigidity, as elaborated by Sherrington and others, can readily be obtained by transection at the level of the anterior colliculi. The animal experimented on assumes a bilateral posture of rigidity in extension, which in Sherrington's view is a close approximation to "reflex standing." All we need bear in mind at present is that when the corticospinal or new motor system is bilaterally and completely out of action, motor reactions, characterized by the adoption and temporary fixity of posture, come into being, or, rather, reveal their presence, the enduring existence of which is under normal conditions largely obscured by the changing phasic activities of the new system. To demonstrate them, the latter has as a rule to be *hors de concours*. In order that they may be thus revealed, mesencephalon, cerebellum, and spinal cord are left, yet the cerebellum is not essential for the reactions. Further, the corpus striatum itself is out of action—a point to which attention is particularly directed. In the neuraxis (as constituted by mesencephalon, pons, medulla, and cord) resides a postural motor activity which, if released by experimentation

or by disease at higher levels, will show itself in phenomena of the kind that I have been describing.

Unfortunately, we are not yet in a position to determine to what extent the phenomena of experimental decerebrate rigidity are due to release from *corticospinal* control, and how much to release from *striatal* control. We cannot for that matter at present be sure whether the action of the corpus striatum or of part of it—the neostriatum—is in reality one of control or inhibition of lower motor centres. The evidence furnished by disease, as we shall see, is suggestive, though not as yet conclusive. In a recent paper Bielschowsky⁵ has assumed for the rigidity of decerebration a component of striatal origin, in the sense that since striatal lesions are associated with rigidity, the removal of that organ in the process of decerebration should add an element of rigidity to that otherwise produced by the experimental lesion. This view is in my opinion open to objection, for the rigidity of decerebration is a specific extensor or flexor-extensor rigidity quite other than the generalized skeletal hypertonia of striatal disorder.

Taking, however, the neuraxis as “stripped” by experimentation, and extending from mesencephalon to cord, inclusive of the cerebellum, we cannot in the present state of knowledge assign the postural phenomena to a mechanism activated after one fashion only. There is a mesencephalic component, a labyrinthine component, and, indeed, a cutaneous component. Otherwise expressed, in decerebration the anatomophysiological system concerned with posture is free to assert itself through stimuli from several different sources. In response to passive alterations of the position of the head in space, or relatively to the trunk, the decerebrate animal makes movements of the limbs and trunk which are the result of influences reaching spinal centres from the semicircular canals, via the pons, or from cutaneous end-organs in the neck, via the medulla (the “tonic-labyrinthine” and “tonic-neck” reflexes of Magnus and de Kleijn). We have seen, too, that postural movements of the limbs can be readily evoked in the same animal at levels definitely above those of the vestibular system, viz. from the mesencephalon. As for any part played by the cerebellum, the recent work of Pollock and Davis⁶ goes to show that one of its functions is to *inhibit* rather than facilitate postural activities, a view that receives some support from the researches of Rademaker.⁷

In a word, from or through thalamus, mesencephalon, pons and medulla, movements and attitudes can be mediated, but *not* from

corpus striatum. We have seen they are postural rather than phasic activities. Even when they approximate to phasic action, as in stepping or progression movements, they never take place with the freedom and rapidity of corticospinal movements.

It cannot be too definitely urged that this assumption of attitude in rigidity takes place at the spinal cord level. When old and new systems, the former in its prespinal portion, are both out of action, as in the spinal animal of the experimentalist, numerous motor spinal integrations are still possible—such as stepping movements, scratch reflex, extensor thrust, and so on. At the bidding of appropriate stimuli what remains of the spinal cord can carry out reflexes and automatisms, sometimes of considerable complexity. The main feature of these experimental automatisms is their invariability; they exhibit a fixity or organization of response that is patently far removed from the “fluidity” of corticospinal innervation.

Our summary may be somewhat in the following terms: (1) We may conceive of the spinal cord as constituting the veritable oldest motor system, and as still capable, in complete isolation, of living and functioning, of exhibiting integrative combinations of movement in a more or less fixed and organized fashion. The patterns of these movements are as it were stamped on the cord from the earliest days in phylogenetic history; the tone they manifest depends on the intactness of proprioceptive arcs for various segmental levels of the cord. (2) These primitive spinal integrations can, however, be activated from a higher level, viz. that of mesencephalopontine centres. When these, in their turn, are completely isolated from above but remain in connexion with the spinal neuraxis, then specific postures can readily be obtained, e.g. efforts at reflex standing; for the different segments of the body (head, trunk, limbs) postures are obtainable which are bilaterally symmetrical and which under suitable circumstances can be brought into action unilaterally. Various spinal units are “re-represented” at this second level, in the sense that they can be combined from it; the leading-reins can be handled by centres situated above the cord but below cortex and basal ganglia. From mesencephalon and from pons these spinal units can be made to act, in various permutations and combinations, yet, as before, the types of response are more or less fixed and organized. (3) The crucial experiments of separation at a third level, viz. that which physiologically isolates both corpus striatum and optic thalamus

from the pallium, while they remain in connexion with the levels below, are still largely awaiting (Goltz's old experiments being in this respect unsatisfactory, while in the "thalamus animal" of Magnus the corpus striatum is removed), and therefore discussion here at once becomes speculative. It is none the less probable that the spinal neuraxis can once more be played on from above, certainly from the optic thalamus, but not, apparently, from the corpus striatum. The fact that movement can be elicited experimentally from cerebellum (according to Horsley * and others), mesencephalon, and optic thalamus, respectively—all via extrapyramidal paths—but not from corpus striatum, might be taken to suggest that the relation of the latter organ to the others may be one of superiority, conceivably on the analogy of frontal (psychomotor) in distinction from rolandic (motor) activity, as already suggested. However this may be, the corpus striatum seems to stand in a different position from the others; it appears to control innervation but not to originate it, just as one of its component functions is tone-control. As transcortical inhibition may fail, allowing involuntary cortical movement (see page 232), so may striatal inhibition fail, and extrastriatal involuntary movement ensue.

(4) As for the influence of the cortex on all this complex potentiality of movement at mesencephalic and spinal levels, we fortunately possess some experiments of Graham Brown ³ which are in my opinion of the first importance and significance. In the decerebrate ape Graham Brown produced the usual mesencephalic reactions already discussed, say postural flexion of the left upper extremity. While this was persisting, the appropriate crus (corticospinal tract) was stimulated. Immediate augmentation of flexion occurred; after withdrawal of this crus stimulus, one might expect the mesencephalic flexor after-discharge to remain unimpaired. But this is not the case. On the contrary, the postural after-discharge vanishes immediately, just as after an ordinary pyramidal stimulation the arm "flops." In other words, stimulation of the corticospinal system "wipes out" the existing postural reaction derived from stimulation of the non-pyramidal system. As Graham Brown says, "non-postural cerebral activity seems to abolish postural midbrain activity."

* Horsley never published the actual details of his excitation of the cerebellum in collaboration with R. H. Clarke. From personal knowledge, however, I can state that he obtained torticollis-like movements of the head by stimulating the dentate nucleus electrically.

The significance of this will at once be obvious to the reader. At any moment cortical, "voluntary" activity obliterates that of lower motor centres; rapid, phasic changes are possible because of the master control of the cortex, the actual nature of the activity of which has in this respect now received physiological demonstration.

Clinico-Anatomical Investigation

Our approach from the clinical side should be without prepossession, and should give due weight to the findings of pathology. We should not allow ourselves to be unduly influenced by what we have learned from the foregoing brief sketch of some of the doctrines of comparative anatomy and some of the data of experimental physiology.

Still, we cannot be far wrong in holding to the double type of motor reaction in man—the phasic, modifiable activities of the corticospinal system, and the automatic, postural, static, more organized and less modifiable activities of the older motor system.

To begin with, then, it must be evident that in health both old and new motor systems are in action. The activity of one does not mutually exclude the activity of the other. If possibly some of the results of experiment appear to obscure this issue, let us be quite clear in the matter. If decerebration lays bare the functions of the old motor system as distinct from the other, it does not follow that in health there is not a fusion of the respective functions of the two. On the contrary, the obvious point has been emphasized by Strümpell⁹ that an indispensable preliminary to, and accompaniment of, conscious innervation is functional activity on the part of the static or postural mechanism. Normally, there is "neural balance" between the two. In the words of Mourgue,⁸ the apparatus for the autoregulation of attitude must be in being if cortical excitations are to effect movements and acts. Winkler¹⁰ expresses the same idea when he says that with each displacement of the head a given attitude of the whole body is determined, and it follows that for each intended "voluntary" movement the body finds itself, reflexly or automatically, in such a position as to enable the appropriate contraction of the muscles to be attained at the moment of production of that "voluntary" movement. Simple as this may appear in theory, the mechanism for tonic autoregulation is none the less of considerable complexity, and its analysis in disease or, rather, the separation in diseased conditions of the activities of the different motor components is peculiarly difficult.

DISEASE OF THE CORTICOSPINAL SYSTEM

In an ordinary hemiplegia, familiar to every clinician, a destructive lesion of the pyramidal tract gives rise to a particular clinical picture. However *unspecific* the pathological condition—for example, a hæmorrhage implicating the internal capsule—as long as it is adequately inhibitory of corticospinal function the arm and the leg on the affected side assume a *specific* “pattern.” This simple fact is in itself surprising enough. The clinical result is the predilection-type, as it is sometimes called, of Wernicke-Mann—in the arm, adduction at shoulder, flexion at elbow, semipronation of forearm, flexion at wrist, flexion of fingers; by comparison, the pattern in the leg is one of relative extension, with some adduction at the hip, and with foot extended and inverted, and toes pointing down and in.

With unimportant variations and few (though none the less interesting) exceptions, this specific attitude of the contralateral limbs follows an unspecific interruption of the corticospinal or new motor system. It is impossible to get away from the fact that disease of that system releases an action-pattern innate in the spinal cord, a physiological grouping, according to a specific scheme, of collections of lower motor neurone units. Into this pattern tone flows, also as a sequel to the lesion, so that the attitude is maintained and only with great difficulty overcome or modified by forcible innervation with what remains of the new motor system. A postural state has taken the place of a phasic state.

The meaning of this precise posture has given rise to a great deal of controversy, which cannot be entered on here, though I may for the moment allude to the view which sees in the hemiplegic attitude a regression to the climbing habits of our postulated animal ancestors, and refer the reader to the excellent criticism of this hypothesis by Mourgue.⁸ The careful investigations of Kraus and Rabiner¹¹ have resulted in the establishment of a formula for these patterns based on a consideration of the anatomical position (dorsal or ventral) of the muscles involved. They have shown that the formula for the extensor attitude of the leg—an antigravity posture—is *VDV*, i.e. ventral muscles are in contraction at the hip, dorsal at the knee, and ventral at the ankle. For the upper limb, however, the formula that corresponds is not found in hemiplegia, but in the extension-pronation posture which I have described as frequently occurring in decerebrate rigidity in man. Here for the

arm it is also *VDV*, an antigravity attitude. Its corresponding opposite, a flexion attitude, *DVD*, is not quite identical with the flexion attitude of the hemiplegic arm, since in the case of the latter the wrist is commonly flexed, as in the decerebrate posture, whereas in the exact opposite we should have extension.

The positions assumed involuntarily by the lower extremities in cases of paraplegia are similarly specific, however diffuse the lesions may be. We distinguish clinically a paraplegia in extension and a paraplegia in flexion. The former is common and familiar; the latter, less common, is as a rule encountered when increasing isolation of the lower section of the spinal cord takes place from spread of the lesion, amounting to a total transverse interruption of function. In addition, we note how in paraplegia involuntary movements of a spasmodic nature are prone to occur. We recognize involuntary flexor and involuntary extensor spasms, the legs drawing up spasmodically, or shooting out straight. Again, we have occasional opportunity to observe how involuntary flexion of one leg is accompanied or succeeded by involuntary extension of the other. We know the various clinical procedures (e.g. that of Marie-Foix) employed to demonstrate these involuntary activities. Instead of describing them as defence reflexes, it conduces to clarity of conception if we regard them as more or less ineffectual attempts at reflex stepping or walking.

Briefly, in these released action-patterns resident in the spinal cord we see evidence of activity of the old motor system in proportion as the new is interrupted; they are not modifiable in the ordinary sense, but are on the contrary integrated according to fixed schemes. As already indicated, they are usually accompanied in action by an access of tonus, so that postures are maintained and involuntary movements are slow and spastic, with lingering after-discharge.

I do not doubt that the isolated spinal cord is the seat of still other reflex motor integrations than those concerned with stepping. Disease brings out such integrations in respect of other physiological groups. In a case of total transverse lesion I found that cutaneous stimulation of the skin area including the external genitalia was always followed by tonic, lingering contraction of the toes in plantar flexion on both sides. The possible biological significance of this and other analogous reflexes that might be mentioned will doubtless suggest itself to the mind.

The question of the hypertonus accompanying these spinal

phenomena is a study in itself. When the lesion is anatomically complete, tone depends on the integrity of the individual proprioceptive arcs of the segments that remain. It is impossible that any of the various other mechanisms associated with tonus control—labyrinthine, mesencephalic, striatal, or (possibly) cerebellar—should be here in action, since they are anatomically isolated.

DISEASE OF THE OLD (STRIATOSPINAL) MOTOR SYSTEM

Thanks to the labours of a number of workers belonging to different schools, English and foreign, the last decade has seen the introduction of order into a formerly very confused subject. The credit is due more to the well-tried clinicopathological method than to any other. As a consequence, we now know that certain diseases are with greater or less definiteness to be assigned to the corpus striatum and its projection system. For our present purpose I need only specify progressive lenticular degeneration, paralysis agitans, and torsion spasm. A syndrome of the corpus striatum has been established by the Vogts, and independently and a little differently by myself, and still more recent work tends largely to confirm the outlines already sketched.

The cardinal clinical features of disease of the corpus striatum may be summed up in three words—variability in muscle tonus, the appearance of involuntary movements, and the absence of paralysis in the strict sense of the term (see also page 143).

Muscle Tonus.—To take the matter of muscle tone first: In the great majority of cases of striatal (extrapyramidal) disease there is permanent increase of tone in the skeletal musculature; in a minority, the tone is variable from time to time, and may therefore be usefully described as constituting a condition of dystonia. A prominent characteristic of cases of progressive lenticular degeneration is the rigidity of the musculature generally, not merely of the limbs and trunk, but also of the face, throat, larynx, etc. It is of fundamental importance that we should remember that this hyper-tonicity does not seem to single out special groups, as flexors or extensors, but affects all groups more or less equally. When the forearm is passively flexed at the elbow resistance is encountered in the extensors, and vice versa when the forearm is extended. Now we have seen that in corticospinal (pyramidal) disease there is a *selective* accession of tone, as in the flexed arm of hemiplegia, the extended or flexed leg in paraplegia. Here, then, we have one

means of differentiating pyramidal and extrapyramidal conditions. Similar phenomena are highly characteristic of paralysis agitans and, more especially, of the remarkable number of postencephalitic Parkinsonian cases encountered by the clinician during the last few years, some of which are truly phenomenal in their generalized rigidity. I regard it as of significance to note that no amount of bilateral corticospinal motor disease can produce hypertonus in all muscles indifferently; in double hemiplegia the tone-picture of postencephalitic Parkinsonism is *not* obtained in its entirety by any means. It is true that in the latter condition there is a general attitude of moderate flexion, which means only that flexor tonus rather predominates, yet there are other instances of the disease, both idiopathic and postencephalitic, as is not well enough known, in which the back is much more erect, in fact slightly extended.

Of equal interest, if much less frequent, is the condition now usually called torsion spasm. In it, temporary hypertonus follows temporary hypotonus in a confused and irregular fashion; the limbs at one moment are contracted involuntarily in flexion, at another in extension, with varying tonic maintenance. As yet only two cases have been examined pathologically (Thomalla,¹² Wimmer¹³) and in each cirrhosis of the liver was found, coupled with changes in the corpus striatum, but also equally marked elsewhere throughout the brain, including the cerebellum, especially in the case recorded by Wimmer. They do not therefore afford the same clear-cut picture as some other forms of striatal affection.

Involuntary Movements.—In respect of the involuntary movements of lesions of the old motor system, those that accompany striatal disease may be classified as belonging to the group either of tremors or of choreo-athetosis. Without doubt, and in spite of the assertions of some continental observers, tremor is much the more frequent of the two as far as the corpus striatum and its projection system are concerned. Only in a minority is choreo-athetosis met with.

It is essential that we should recognize the full import of the fact that the striatal lesions with which hypertonicity and involuntary movements are associated are destructive lesions. Whether there be a general disintegration and necrobiosis of the organ, bilaterally, as in progressive lenticular degeneration, or a more or less chronic outfall of cells and fibre-systems, as in paralysis agitans, or an interruption of neuronie activity from a fibre overgrowth, as in the status marmoratus described by the Vogts, each and all

are negative or destroying lesions, and it must follow that the mechanisms for the involuntary movements and the hypertonia or dystonia are nonstriatal. This conception has been familiar to the English neurologist since the days of Hughlings Jackson, but it has only recently begun to receive widespread recognition. In their recent communications, for example, the Vogts¹⁴ are compelled to admit that the hyperkineses resulting from disease of the corpus striatum are of "substriatal" origin. "Extrastriatal" or "nonstriatal" is preferable to "substriatal." They make no attempt, however, to specify or localize the mechanisms more particularly.

This clinicopathological fact bears out, it will be seen, the views already adumbrated above, that the relation of the corpus striatum to the rest of the old motor system is one of tone control, and of control of innervation. Remove its influence by disease, and cerebello-mesencephalo-spinal motor mechanisms come into overaction *in spite of* the normal activity of the pyramidal system; tonic postures become overemphasized; in fact, a universal muscular rigidity appears, distinct from the selective rigidity of corticospinal disease; or, alternatively, involuntary movements develop, over which the control of the master system, the cortico-spinal, is at best fleeting and imperfect.

I wish to draw special attention to the fact that the normal corticospinal system is unable to prevent the effects of striatal disease from making their appearance. Though we speak of it as the "last word" in motor control, it cannot inhibit the hypertonia and hyperkinesis of striatal disease. I believe we have here proof of the relatively autonomous nature of the corpus striatum, which is to be expected from the fact that practically no corticifugal fibres enter it.* The evidence I have adduced indicates how independent, in spite of its phylogenetic age, is its influence on the spinal motor centres.

Efforts have also been made by the Vogts to distinguish clinically lesions of the neostriatum from lesions of the palæostriatum, in an endeavour to bring the symptomatology of dystonia and hyper-

* The question of corticopetal or corticifugal fibre connexions with the corpus striatum is highly controversial. I have stated my conclusions, as far as the ape is concerned, in the paper already alluded to.¹⁵ Recent work by Minkowski,¹⁶ Kodama,¹⁷ and others, would appear to show that the ganglion is not quite so isolated from the cortex as has been thought. Coenen¹⁸ finds no connexion between cortex and putamen in rabbits, but some between globus pallidus and both temporal and rolandic regions.

kinesis into a striatal scheme. They maintain that lesions of the putamen-caudate (neostriatum) are associated with involuntary movements (tremor, chorea, athetosis), and of the globus pallidus (palæostriatum) with rigidity. Bielschowsky, similarly, gives some support to this view. Ingenious as is the scheme, it is open to objections, only some of which can here be mentioned. (1) Following Hughlings Jackson, I believe that tremor and rigidity are sometimes closely interrelated. Now in progressive lenticular degeneration, tremor and rigidity are both early symptoms, and the earliest lesion is without question in the putamen-caudate, the globus pallidus being intact. (2) Bielschowsky says that destruction of the globus pallidus produces a rigidity which masks the involuntary movements caused by implication of the putamen-caudate. Yet in cases in which *both* parts of the corpus striatum have been destroyed tremor may continue to form a very prominent symptom. (3) Tremor and choreo-athetosis are involuntary movements very different in type, and it is inconceivable that the same mechanism should produce both (see page 210). (4) Numerous cases are on record in which lesions determining the appearance clinically of choreo-athetosis or of tremor have not been situated in any part of the corpus striatum. To speak, as the Vogts are compelled to do, of a syndrome of the corpus striatum of "thalamic origin," of "cerebellar origin," etc., is both confusing and unjustifiable.

We must, no doubt, accept the apparent occasional occurrence of choreo-athetotic movements from lesions of the corpus striatum; they have been observed in a few cases of progressive lenticular degeneration and in cases of status marmoratus; nor in this respect must we forget the apparent association of hyperkineses of this kind with striatal lesions as in torsion spasm, or the seemingly definite lesions of the same ganglion in Huntington's chorea (cf. Dunlap¹⁹).

I²⁰ have, however, repeatedly emphasized the importance of realizing the facts of the occurrence of choreo-athetosis and of tremor in lesions not situated in the corpus striatum; as regards the former, it has been seen in lesions of the optic thalamus, regio subthalamica, superior cerebellar peduncle (Bindearm-chorea), etc. I have elsewhere advanced arguments (see page 230) to show that lesions associated with its occurrence are found mainly on the afferent cerebello-mesencephalo-thalamo-cortical path, and that the corticospinal tracts must be, comparatively speaking, intact. If they are thrown out of action completely, by disease, the involun-

tary movements cease; significantly, neither in decerebrate nor in thalamus animals (Magnus) are such movements found. In both instances pyramidal function is nil. Horsley²¹ checked persistent athetosis in an upper limb by excision of the corresponding motor cortex area. Experimental researches by Lafora²² (though they require confirmation) favour the view here once more outlined. He has produced persistent choreo-athetoid movements in cats by lesions of the cerebellar peduncle, mesencephalon, and regio subthalamica.

The problem of the apparent association of *some* striatal lesions with the same clinical phenomena remains for solution. I suggested a number of years ago that some intimate connexion by striothalamic fibres, of which there are large numbers, might conceivably explain the facts (if they are facts). The play of striatal impulses on the optic thalamus may resemble that of the cerebello-mesencephalo-thalamic system, in the sense that both are afferent or thalamipetal, and that in circumstances not yet elucidated lesions on either path may occasion choreo-athetoid movement via the corticospinal system (see also Chapter X).

The case of tremor is somewhat different. Its association with midbrain and tegmental lesions generally is undoubted, and I have on previous occasions laid stress on the absence of any evidence to implicate the cortex in its production. Hughlings Jackson declared that "tremor differs from rigidity, not fundamentally, but in degree," and I have always maintained that herein lies the clue at least to *some* of its appearances. Without here going further into this question also, I may say that I associate tremor with the effect both of striatal and of cerebellar impulses on the mesencephalon, and that in the case of the former tremor is more prone to be associated with exhibitions of rigidity than in the latter; indeed, with the latter the tonus of the muscles involved may be seemingly diminished.

The reader is referred to Chapters X and XI for a more detailed examination of the problems presented by involuntary movements.

SIMULTANEOUS INVOLVEMENT OF CORTICOSPINAL AND STRIATAL MOTOR PROJECTION SYSTEMS

The compounding of lesions of old and new motor systems is the last matter I shall touch on. We have seen that in normal circumstances a constant interplay of old and new motor activities must occur. In diseased conditions in man we rarely get anything

like complete outfall of function of one or the other system. Disease has, however, shown that in lesions of the corticospinal paths paralysis of voluntary movement is associated with the assumption of involuntary postures, produced by the release of function of spinal motor units in action-patterns which must have some significance. Disease of the striospinal system, on the contrary, "allows" rigidity and hyperkinesia. The rigidity is much more widespread than in corticospinal cases, influx of tone affecting flexor and extensor action-patterns more or less equally and simultaneously, with the clinical result already described.

Complete Interruption of Function.—In the condition which has been regarded as decerebrate rigidity in man, i.e. one of complete interruption of function—both corticospinal and striospinal—produced by lesions of various kinds roughly speaking at the level of the mesencephalon, what do we find? Since the striatal system is out of action, we might expect the universal rigidity of the postencephalic Parkinsonian, but it does not develop. Instead we get an attitude in extension which may fairly be regarded as indicative of an attempt at "reflex standing." In other words, the posture produced by release of function when both old and new systems cease to act on the spinal neuraxis simultaneously is that of release neither from the one nor from the other.

Decerebrate attitude is a pattern-attitude, as is the case with corticospinal disease, but it is neither that of double hemiplegia nor of paralysis agitans. Into this innervation-pattern tone flows, yet there is no widespread, apparently non-specific overflow, as seems to be characteristic of striospinal disease. The explanation is, possibly, that the effect of release from cortical motor control outweighs the effect of release from striatal control.

We have seen that experimentally cerebral activity can be shown to abolish midbrain activity; its control over the latter exceeds that exercised by the corpus striatum, we may suppose, and when both systems are out of action the clinical result approximates rather to that which follows release from the former than to that consecutive to release from the latter. It is certain, in any case, that the effect of the double release is not a mere arithmetical summation of the two single release-effects, and we are compelled to admit that the consequence of compounding the two, if they are complete, is not quite what might be expected.

An entirely analogous problem has to be faced by the physiologist. Magnus²³ has shown that when only the pyramidal tracts are cut

in the intact animal little abiding alteration of tone develops, but such influence as they exert is flexor in character. When the rubro-spinal connexions (at Forel's decussation) are cut, also in the intact animal, there is only slight rigidity. But when the latter are sectioned in the thalamus animal (pyramidal tracts already cut) *maximal* extensor rigidity ensues. Thus the *compounding* of the two influences, released by section, is distinctly different from what would appear to be the *summation* of the two considered separately. Herein lies one more of the problems still calling for solution.

Incomplete Interruption of Function.—When the two systems, corticospinal and striospinal, are each incompletely out of action, we shall naturally find a complexity of symptoms difficult to unravel. Take a case of infantile cerebral hemiplegia with athetosis, and with some voluntary movement of the affected limbs still possible. Partial abrogation of corticospinal function causes the hemiplegic attitude to appear; this in its turn is constantly being modified by the appearance of involuntary movements, which for the sake of argument we may suppose of extrastriatal origin. The result is a medley of paresis, hyperkinesis, and dystonia which we might almost despair of analysing.

I am convinced, none the less, that by grasping the principles of corticospinal and striospinal action, as I have tried to indicate them, by studying their effects in pure and uncomplicated cases exhibiting the results of disease of the one and the other separately, we shall eventually reach a position from which we shall be able to solve some outstanding difficulties concerned with the human motor system in health and in disease. The clinician and the neuropathologist must contribute their quota to the attainment of this desirable end just as much as the experimental physiologist.

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- ²⁰ WILSON, KINNIER, In *Questions Neurologiques d'Actualité*, 1922, p. 1.
- ²¹ HORSLEY, V., *Brit. Med. Jour.*, 1909, ii, 125.
- ²² LAFORA, R., *Asoc. Espan. Prog. Cienc.*, Oporto Congress, June, 1921, 207.
- ²³ MAGNUS, R., *Körperstellung*, 1924, 639.

CHAPTER VII

DISORDERS OF MOTILITY AND OF MUSCLE TONE, WITH SPECIAL REFERENCE TO THE CORPUS STRIATUM *

I. *THE VOLUNTARY MOTOR SYSTEM IN STRIATAL DISEASE.*

Introduction. The Voluntary Motor System. Classification of Muscles in a Normal Act. Voluntary Movement in Striatal Disease. Strength or Weakness of Movement. Micrographia. Beginning, Course, and Cessation of Movement. Poverty of Movement. Causes of Parkinsonian Akinesis. Akathisia.

The rise into prominence of problems connected with the functions and semiology of the corpus striatum forms perhaps the most striking feature of contemporary neurology. At first contributed to by but one or two workers abroad and at home, the study of this part of the brain has been vastly augmented during the last decade and is already responsible for an unbelievably extensive literature. Notwithstanding this immense activity, finality is far from being reached in respect of both the pathology and the physiopathology of symptoms attributed to its affections, and speculation is correspondingly rife. Motor symptoms as widely divergent as the complexity of an elaborate tic and the simplicity of a single muscle twitch or myoclonic contraction, as heterogeneous as palilalia and micrographia, are alike set down to striatal disease. Now the corpus striatum is so situated that traumatic lesions are not calculated to produce any clear-cut picture of its symptomatology; its vascular supply is not derived from a single source; morbid processes often involving its cell- and fibre-systems (e.g. that of epidemic encephalitis) are not restricted to the ganglia in

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question ; the immediate vicinity of the corticospinal tracts sometimes renders it uncertain, in disintegration cases, whether there is not also concomitant interference with the function of these ; even in Huntington's chorea, confidently regarded as a striatal affection because of the outfall of cells in the corpus striatum found with some frequency, every one knows that the cerebral cortex is also simultaneously implicated.

For these and other reasons the ganglia situated in the base of the brain still, to a large extent, retain the characteristic of basements—viz. darkness. Since hypotheses erected on an uncertain pathology have outstripped clinical analysis, I have undertaken an objective investigation of various symptoms of a motor order assigned by many, on insufficient or erroneous grounds, as I shall endeavour to prove, to striatal disease. The title has been chosen to avoid giving the impression that the clinical phenomena to be considered are in my view and without comment to be set down to striatal disease and to that alone.

The Syndrome of the Corpus Striatum

In 1912, as a result of work ¹ on the disease to which the name of "progressive lenticular degeneration" was given, the following syndrome of the corpus striatum was put forward :

"In pure, uncomplicated, bilateral lesions of the lenticular nucleus, and more generally of the corpus striatum, provided they are of sufficient size and of adequate duration, the clinical symptoms are bilateral involuntary movements, practically always of the tremor variety ; weakness, spasticity, or hypertonicity (sometimes spasmodic contractions), and eventually contracture of the skeletal musculature ; dysarthria or anarthria and dysphagia, and a degree of emotionalism ; but without any sensory disturbance, without any true paralysis, and without any alteration in the cutaneous reflexes. If the abdominal reflexes are absent (apart from muscular rigidity), or the plantars of extensor type, then the syndrome is no longer pure."

While subsequent research has fully confirmed the general accuracy of this description, further investigation has filled in certain gaps then left more or less open, while in other respects, as we shall see, a more ingoing analysis of the symptoms then specified is now required.

In the same monograph the considerable resemblance between progressive lenticular degeneration and paralysis agitans was more than once alluded to, and approximate similarity of the pathology of the two conditions was reasonably surmised. This expectation

is now known to have been correct, the essential pathology of Parkinson's disease consisting in degeneration of certain striatal and striofugal systems. Since progressive lenticular degeneration is comparatively rare, while Parkinson's disease and the Parkinsonian syndrome are common, we may take the latter as a working example of a mainly striatal malady. Striatal motor symptomatology may be summed up in three words : variability in muscle tone (dystonia), the appearance of "involuntary" movements, and the seeming absence of "true paralysis." As the last of these requires qualification and explanation, we may begin our study with it.

The Voluntary Motor System

The term "voluntary," applied to movements, is in constant employment, but of elusive connotation. "Voluntary" movements are not *sui generis* in the sense of constituting a class apart ; they are equivalent to "least automatic" movements, and all gradations may occur from "most automatic" to "least automatic." In objective characters they are not to be distinguished from other types of movement, equally co-ordinated and apparently purposeful, from which the element of volition is nevertheless wanting. No single criterion of an objective clinical kind (by string-galvanometer methods or otherwise) leads to differentiation ; the will makes a subjective though not an objective contribution to a movement. Ordinarily speaking, we are in the habit of regarding as voluntary those movements which we have reason to believe are effected through corticospinal or pyramidal tracts, to some extent because in the experimental animal movements (not, as a fact, strictly comparable to those of volition) can be produced electrically via these tracts. The falsity of this deduction is apparent when we remember the ease with which movements can be electrically elicited from other paths never considered "volitional."

Consciously, we are in ignorance of the specific muscular units effecting a voluntary movement. Consciousness knows only movements and terminal joint displacements produced by movement, and this has sometimes been taken to argue the representation of movements only in the cerebral convolutions. Evidence will be adduced later which, I believe, disproves the contention. The point at present is, that a large part of every voluntary movement is both involuntary and outside consciousness. The expression "voluntary muscular contraction" must therefore be regarded more as a *façon de parler* than a term with distinctive quality.

Similar difficulties follow in respect of the term "paralysis," which is equally vague and open to misinterpretation. The clinician admits degrees of paralysis, but would have hesitation in defining with precision exactly what they signify. In a given voluntary movement the elements of power of muscular contraction, appropriate co-ordination of muscular units, rate of contraction, accuracy of timing, estimation of amount of contraction required, are among some of the factors in the attainment of a perfect end, and if in any of these the voluntary movement falls short it exhibits in that respect a degree of paralysis or paresis. If a movement is normal except for its unusual slowness of accomplishment there is paralysis of normal rapidity of contraction. When, in the definition of the striatal syndrome cited above, mention was made of the "absence of true paralysis," the endeavour was to separate paralysis ensuing on lesions of the pyramidal system from the "motor helplessness" of extrapyramidal cases, in which, as was shown, objective signs of involvement of pyramidal paths (absence of abdominal reflexes, presence of Babinski plantar responses) were wanting. It was then suggested that the discovery of an appropriate expression for extrapyramidal "paralysis" was a desideratum.

As it is, such terms as weakness, paralysis, paresis, and even such apparently more precise expressions as "direct weakness of the motor impulse to the agonists" (Foerster³), have, in fact, little if any specificity, however convenient they may be. If the word "paralysis" is indispensable, it may be justifiably employed to indicate imperfection of movement in respect of *any* of the components of a normal movement, and cannot be confined to questions of muscular strength or power alone.

It became apparent to me some time ago that in order to determine with certainty whether any particular kinds of movement were as a fact impaired or lost in disease of the corpus striatum a complete examination of the motor phenomena from the point of view of normal muscular action of various types would have to be undertaken. The reader of striatal literature finds himself confronted by statements as to loss of movements which in the absence of definitions or expositions are exceptionally confusing. Thus he will be told there is, *inter alia*, "loss of primary automatisms" (C. and O. Vogt²), "loss of associated movements" (Foerster³), "loss of individual movements" (Jakob⁴), "loss of the associated movements that accompany voluntary movements" (Sternitz⁵), "loss of primitive automatisms and of higher co-ordinations"

(Binswanger ⁶), "loss of (palæokinetic) automatic and associate types of movement" (Ramsay Hunt ⁷), and so forth. Accordingly, a scheme of examination has been devised and applied to a large number of cases, mainly of the Parkinson group, but including also a typical case of progressive lenticular degeneration and a number of choreics of the Sydenham, Huntington, and senile varieties, as well as cases of athetosis. The terms used will be explained as they occur.

Classification of Muscles in a Normal Act

A physiological classification of the various muscles taking part in a simple movement of the type called voluntary divides them into the following groups: (1) Prime movers or agonists (protagonists); (2) synergic muscles, or synergists; (3) fixation muscles, or fixers; (4) antagonists.

This classification differs somewhat from those of Winslow ⁸ and Duchenne, ⁹ and follows the much more recent one of Beevor. ¹⁰ For simplicity's sake it may be modified to include (2) and (3) under the one heading of synergists. Thus in the elementary illustration of the movement for closure of the fist the agonists or prime movers are the flexors of the fingers and thumb; the synergists are the extensors of the wrist, while the antagonists are the extensors of the fingers. Beevor showed that in firm closure of the fingers the triceps and biceps act as fixation muscles when a certain measurable degree of contraction of the agonists is attained, and regarded the fixation group as fulfilling an essential part in all movements in distinction from the synergists, which he defined as "the muscles which are brought into the combination when a prime mover has, by passing over two or more joints, two or more actions of which only one is required; the synergics are the muscles which neutralize these undesired actions." In the above-given example the synergic muscles, the wrist extensors, serve to obviate flexor deviation of the wrist by the contracting long finger flexors, hence in reality their function is one of fixation. It is practicable, therefore, to include as synergists such muscles as contract (invariably or variably) either to fix joints other than that, or those, actively engaged in the particular movement required, or to enable the prime movers to execute their function with a maximum of effect.

Using the attribute "synergic" in this specific way, I do not speak of "synergic movements" or "movement synergias." In a *whole* movement *part* only is synergic; I shall not use the term to

express a complete movement, or a movement concurrent with another. The necessity of this restrictive definition becomes apparent in view of the meaninglessness of any wider application. All the units in a voluntary movement co-operate, even though some may constitute a "co-operation of antagonism." If synergy is synonymous with co-operation it is otiose. There is the more need for terminological clarity at this point in that Tilney,¹¹ whose contributions to a difficult subject deserve respectful consideration, appears inclined to employ the term "synergic unit" to include *all* the muscular components in a given movement—thus eliminating the muscular distinctions drawn above. For the present, however, it will be convenient to keep closely to a classification which has definite merit.

Voluntary Movement in Striatal Disease

(I) STRENGTH OR WEAKNESS OF MOVEMENT

The customary clinical tests of muscular power have been employed in this connexion; the only apparatus I have employed has been the dynamometer, the same instrument always being used, and in two ways—either by a succession of grips first on one side and then on the other, or by alternate right and left grips. Useful information has been obtained by selecting unilateral Parkinsonian cases, care being exercised rigorously to exclude those in which pyramidal function was not considered normal.

For illustration only three cases need be cited.

1. Left-sided unilateral case.

Consecutive.		Alternate.	
R. 150	L. 104	R. 145	L. 100
144	110	140	100
136	100	140	105
134	114	136	102

2. Right-sided unilateral case.

Consecutive.		Alternate.	
R. 82	L. 98	R. 71	L. 104
84	92	62	90
84	90	82	82

3. Bilateral case.

Consecutive.		Alternate.	
R. 72	L. 48	R. 52	L. 40
65	45	65	42
72	38	42	45

From an analysis of many similar records and a comparison with normal cases the disparity between the two sides in unilateral cases is clearly greater than can be accounted for by the natural right- or left-handedness of the individual concerned; and in bilateral cases the reduction of power, as estimated by the dynamometer, is unmistakable. The conclusion is that *relative motor weakness is to be expected in striatal cases of the Parkinsonian type, varying within wide limits but definite enough*. This is in accordance with the experience of James Parkinson¹² himself, who, in his celebrated *Essay on the Shaking Palsy*, mentioned in his definition the occurrence of "lessened muscular power."

Another point brought out by dynamometer records concerns the question of progressive weakening in a series of similar move-

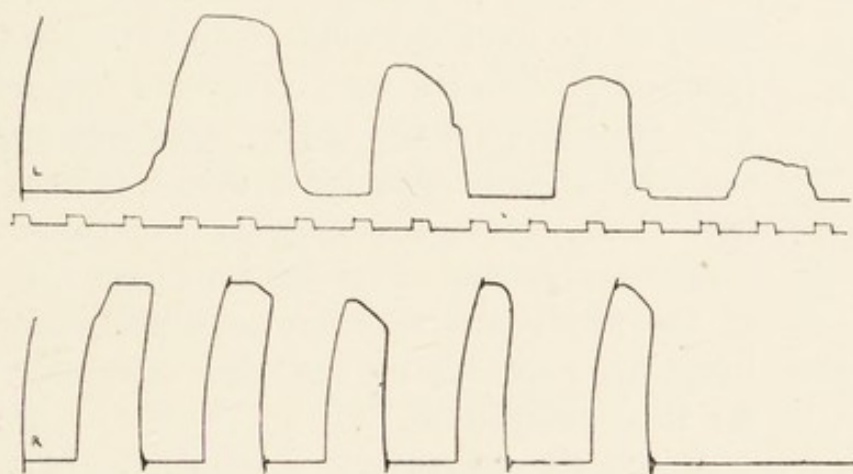


FIG. 5.—Case of unilateral (left) Parkinsonism. Voluntary contractions of right (lower tracing) and left (upper) quadriceps.

ments. This is stated to be a characteristic of striatal affections, but my observations do not substantiate it; at least, not without some qualification. A glance at the above figures will show that in the unilateral instances similar movements (hand-grasping) are at first relatively better sustained on the *affected* than on the unaffected side. Only, however, for a few consecutive movements is this usually found; in a longer series the affected limb undoubtedly tends to weaken more than the other. This is in accordance with the remark of a right-sided Parkinsonian girl, who said: "I could hit anyone hard with the right hand several times quickly, but I couldn't keep it up."

A good way of demonstrating the same phenomenon is by placing the patient in the recumbent position and getting him to make single consecutive contractions of the quadriceps group on the front

of the thigh. The action of gravity is excluded and there is no displacement at the joint concerned, since he is requested not to lift his heel off the couch. From Fig. 5 it will be seen that on the right (normal) side the patient was able to make five successive contractions of the quadriceps in slightly over eleven seconds, the last being as good as the first; on the left (Parkinsonian) side there were four contractions in twelve seconds, and they exhibit a progressive reduction in extent, much more noticeable, however, in the fourth than in the first three.



FIG. 6.—
Case of uni-
lateral (left)
Parkinsonism.
Normal vol-
untary con-
tractions of
right long
flexor of fore-
finger.

Records from a well-marked case of unilateral Parkinsonism furnish evidence of similar weakness in respect of voluntary movements of the long flexor of the forefinger (Figs. 6 and 7). On the right (unaffected) side the tracing reveals free, quick, and wide movement of the finger, three consecutive and smoothly co-ordinated innervations taking place in less than three seconds. By comparison, three similar movements of the left forefinger ("crooking" of the finger) are very slow, extremely limited, and irregular, requiring no less than seventeen seconds for their completion.

Among the movements particularly apt to exhibit weakness are those of separation of the fingers (interosseal). The Parkinsonian cannot readily "spread" his fingers, feebleness and a certain lack of precision being often seen; nor can he approximate the tip of the thumb and that of each of the other fingers in a quick,

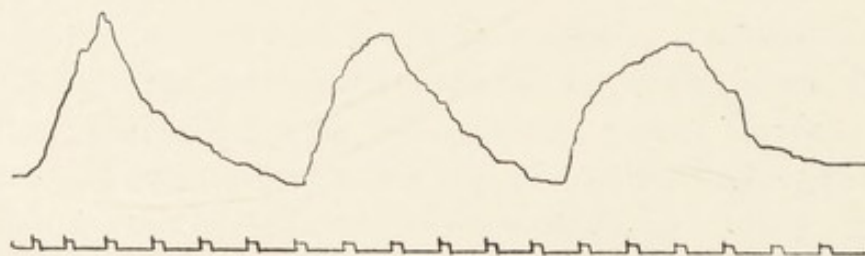


FIG. 7.—Same as Fig. 6. Slow and limited voluntary contractions of left forefinger (affected).

strong, and precise fashion. Abduction of the little finger is not infrequently almost nil. Movements of the tongue, too, both inside and outside the mouth, are especially prone to exhibit the same characteristics. It is protruded imperfectly, and is not infre-

quently bitten in the course of mastication. Speaking generally, the defect is chiefly seen in small-muscle movements, those of larger muscular masses being less obviously imperfect, one reason for which may be simply that gradations of movement are more easily followed by the clinical eye in small than in large muscles, and minor variations of power more easily estimated (see also below).

A further example of this imperfection of volitional movement in the smaller muscles is provided by those concerned in phonation and articulation. A common feature of the phonation process in

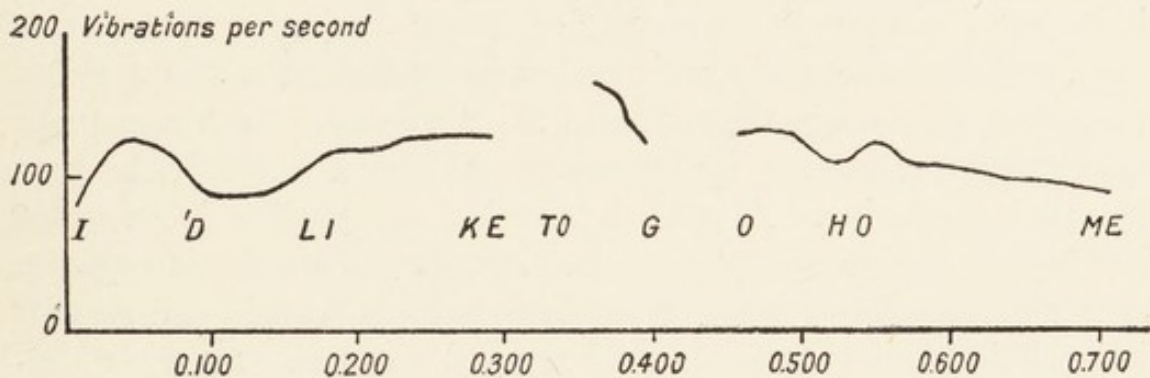


FIG. 8.—See explanation in text.

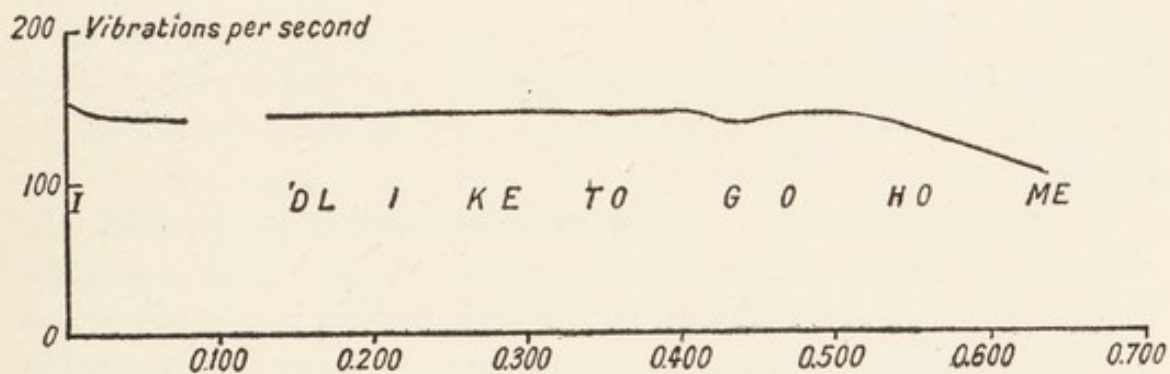


FIG. 9.—See explanation in text.

paralysis agitans, progressive lenticular degeneration, and other striatal syndromes is the monotone voice. I asked Prof. E. W. Scripture to make records for me, one of a normal voice and one of a severe case of Parkinsonism selected for the purpose. The latter, I believe, is the first record of the kind that has been taken. The striking differences between the two can be realized by comparing Figs. 8 and 9. The sentence enunciated in each instance was "I'd like to go home," and while in both the duration was approximately 0.7 seconds, the record of the Parkinsonian is distinguished by complete absence of flexibility, measured in vibrations per second. In other words, here is evidence of imperfection of voluntary move-

ment in a well-defined physiological group of small muscles, revealed by defect of range and of flexibility at a quick rate (that is, the record reveals the presence of muscular rigidity). To dissociate the quotas of these elements may be somewhat difficult, but in so far as they constitute a departure from normal volitional movement of the apparatus in question, they indicate a degree of motor paresis in the sense specified above.

Reference may now be made to the ocular muscles. While as a general rule the external ocular muscles of the Parkinsonian are supposed to exhibit no motor abnormality, in a large number of instances a punctuate movement of lateral conjugate deviation is seen. The eyes move in a somewhat jerky fashion with fractional pauses at intervals in the course of the deviation. Such punctuate movement is, however, by no means confined to cases of the kind we are considering. A more interesting observation is one that has long been a commonplace—viz. defect or failure of convergence. A fair percentage of cases of paralysis agitans (and in this respect it is obviously advisable to deal for the moment only with idiopathic cases) reveal inability to converge, one or other eye usually swinging out, or at least failing to move inwards, at the appropriate moment. A patient told me that one of his earliest symptoms was inability to light his cigar, since he could not with accuracy bring the lighted match to the end of it—a convergence-accommodation defect.

Of its occurrence more than one explanation is possible. In elderly folk the convergence-accommodation mechanism naturally tends to functional involution, but apart from this, its paresis occurs in Parkinsonians who are not old in years and may be observed in postencephalitic Parkinsonism as a solitary ocular defect. Here, evidently, is loss of a particular movement of a set of muscles with preservation of other movements of the same muscles. Hughlings Jackson held there are differences of "evolutionary rank" of movements of the same muscles, and, since convergence-accommodation is a very "special" movement, it may suffer more readily than others. Be this as it may, its frequent impairment provides further proof of feebleness of volitional movement in striatal disorder.

Weakness of closure of the eyes is often encountered, especially in postencephalitic cases. The patient appears unable to "screw the eyes up tightly"; the orbicular muscles usually pass into a state of blepharospasmic trembling. If the eyes are in fact closely shut for a moment or two he not infrequently seems to be unable

to maintain the voluntary contraction tonically. The orbicular muscles of the mouth may act in a similar fashion ; the Parkinsonian tends to have his mouth open or his lips apart, and cannot always "purse them up" strongly, as does the normal individual. If he does, he cannot always maintain the tonic contraction.

I attach no little importance to this *weakness of maintenance of contraction*, which, for that matter, can be demonstrated also in other small muscle movements, and is found (as I have shown) in cases of progressive lenticular degeneration as well. It furnishes proof of the inaccuracy of the common statement that Parkinsonian movement is always characterized by slowness of relaxation. Further, it is a clinical symptom the very opposite of that imagined by some to be typical of extrapyramidal disease—viz. so-called "perseveration" or "tonic innervation." I shall return to this question at a later stage.

As far as movements at large joints are concerned, weakness is much less noticeable. Movements of trunk, head, and proximal limb segments against resistance are sustained with no little force, nor is restriction of range as a rule in evidence, at least contrasted with that of small muscles. I have never seen actual *loss* of movement of a big joint in Parkinsonism such as may be observed in any recent and severe hemiplegia, or, of course, in any paralyzing lesion of the lower motor neurone. If the patient's limb is passively put into a position half-way towards that which he is asked to assume actively, it will often be found to execute the movement with greater power. Thus, if with leg fully extended he is required to flex at the knee, he will accomplish the movement more forcefully should the knee be first passively flexed to a right angle. Foerster³ appears to claim this phenomenon as an alleged peculiarity of striatal affections, but it is in reality a question of mechanical leverage—the adoption of an optimum position no less characteristic of normal musculature than of diseased.

To decide whether in the striatal affections exemplified by paralysis agitans and its congeners any predilection type of paralysis or paresis obtains, as we know is the case in pyramidal affections, we have at our disposal data of the varieties illustrated above, and the general conclusion seems justified that *those muscles whose mass is small are found to exhibit feebleness of voluntary contraction, and that, for the body as a whole, this feebleness is in inverse ratio to the size of the muscle or muscles concerned.* A generalization of this description however, should not be taken too literally or mathe-

matically, though I believe it to be clinically applicable. When a hemiplegic patient slowly and laboriously extends his fingers we do not hesitate to describe the movement as weak or paresed, and no adequate reason can be advanced for refusing the same descriptive epithet to the slow, laborious, and limited spreading of Parkinsonian fingers. Dynamometer records prove the existence of muscular weakness in moderately severe striatal cases, hence the non-specificity of the term "paralysis" already alluded to.

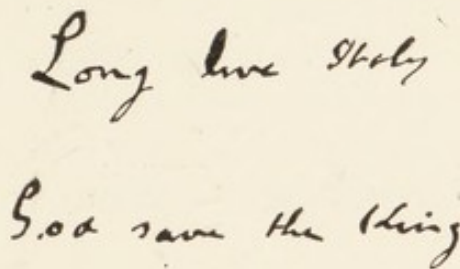
With exception of the ocular musculature—where defect is largely if not entirely confined to the convergence-accommodation mechanism—small muscles in the distributions mentioned above are weak for all movements. They are muscles implicated in execution of the most delicate of acquired movements, especially so in the case of fingers and phonation-articulation apparatus (larynx, tongue, lips); muscles in which fractional gradations of innervation are essential for production of the finest changes, as in speaking and in precise and exact finger movements. This highly special class of acquired movement, then, suffers more than any other in diseases of the group in consideration, and offers in this connexion no essential difference from what obtains in pyramidal cases, except, it may be, in regard to the speech muscles. These are less likely to suffer in hemiplegia, owing to their bilateral innervation. Consonantly with this, unilateral Parkinsonian cases may exhibit little speech—i.e. phonation-articulation—disorder. Loss of highly specialized, recently acquired movements which Hughlings Jackson thought so characteristic of hemiplegia is paralleled by their impairment in Parkinsonian cases.

The cause or causes of the motor impairment exemplified in this section will be more conveniently discussed after certain other features of the voluntary nervous system in striatal disease have been examined.

Micrographia

The behaviour in paralysis agitans of the fine acquired movements of writing may be appropriately considered here. A disturbance of writing known as micrographia, described years ago by Arnold Pick,¹³ but recently alleged to be characteristic of striatal affections by Bernhard,¹⁴ Bing,¹⁵ and others, and indicative of actual paresis by Foerster, deserves remark. The symptom has interested me ever since the appearance of Pick's original paper in 1903 and long before attention was directed more specifically to the corpus stria-

tum. It consists, as its name implies, in an obvious reduction in size of the lettering of the writer in comparison with his calligraphy before development of the organic lesion effecting the change. According to my own observations a distinction may be drawn between micrographia as obvious at the beginning of a sentence as at its close, and that which consists rather in a tailing off into a smaller lettering because of fatigue or other inability to maintain the range voluntarily. In respect of the former variety (to which perhaps the term might be restricted) *it is neither confined to striatal cases nor pathognomonic of them*, and Bernhard commits a *petitio principii* by confining his researches to cases in which striatal disorder has been evident or presumed. It may be observed typically in hemiplegia and other corticospinal conditions. For example, the writing reproduced in Fig. 10 is that of a patient who developed a right brachial monoplegia of cerebral syphilitic origin,



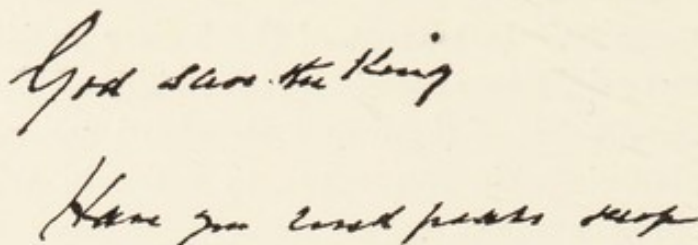
Long live Italy
God save the King

FIG. 10.—Micrographia in cerebral (cortical) neurosyphilis.

from which a good degree of recovery was made. He was a professional mandolin player and complained of two residual symptoms: (1) He had lost all facility in making the fine movements of striking the mandolin strings with his plectrum, though otherwise he could use the arm with ease; (2) since the stroke his handwriting had become unaccountably small in comparison with his previous style. Loss of acquired movements of plectrum-striking constitutes a limb-kinetic apraxia in Liepmann's sense and definitely suggests a cortical origin for the disorder, while the concomitant and otherwise monosymptomatic micrographia can legitimately be assigned in this case to disturbance of cortical function. From a case of cerebral arteriosclerosis the writing seen in Fig. 11 was obtained. It was that of a patient with slight right hemiplegia and defect in speaking, and had been noticed since the onset of right-sided weakness some months previously. No trace existed of the arteriosclerotic muscular rigidity often assigned (by Foerster and others) to striatal disease, nor any symptom of a kind that could be referred

to the ganglion. On the contrary, the evidence pointed to implication of the cortex in the distribution of the left sylvian artery.

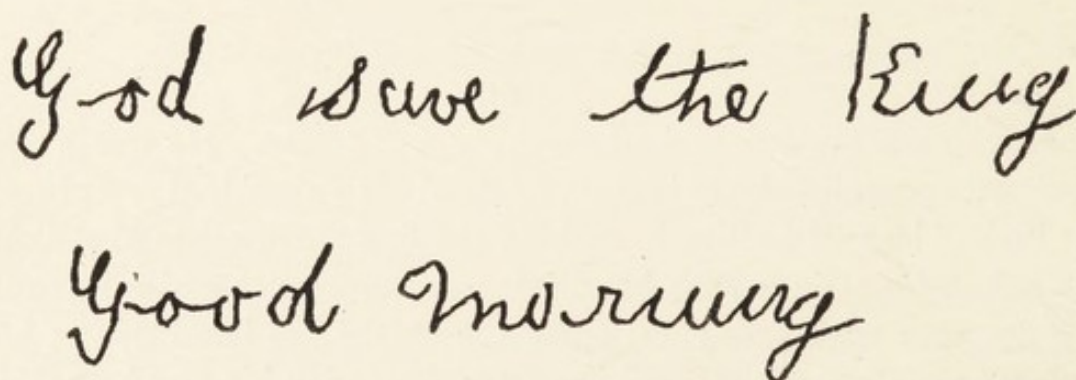
Taking next Parkinsonian cases, I have seen many unaccompanied by any micrographic disorder. The handwriting reproduced in Fig. 12 comes from a typical case of postencephalitic Parkinsonism, the patient being one of the most immobile and rigid I have so far



God save the King
Have you ever heard of a king

FIG. 11.—Micrographia in cerebral arteriosclerosis (non-striatal).

observed. When tested with the dynamometer he did not reveal by a single muscular flicker otherwise the strong effort he was making with his right or left hand, but his calligraphy shows no departure, as far as size goes, from his previous normal. By way of contrast, the most pronounced micrographia in my collection is that of a lady who came under observation only a month or two ago, one of the



God save the King
Good morning

FIG. 12.—Normal handwriting in a case of severe Parkinsonism.

encephalitis cases of the spring of 1924. Her Parkinsonism made its appearance in August of the same year, and her micrographia is such as to render her writing almost illegible (Fig. 13), yet, otherwise considered, the symptom-complex in her particular case is of only moderate degree.

For the understanding of this somewhat involved subject micrographia should not be divorced either from its opposite, macrographia, or from the frequent concomitant of macropsia or micropsia. As far as I have noticed, Bing¹⁵ only of recent writers refers to

these, solely, indeed, to macrographia, which he calls "megalographia," and thinks is of cerebellar origin. He uses the illustration as an argument for the existence of symptomatological opposites between striatal and cerebellar disease. Recent contributors to the subject have failed to take into consideration the variety of cases in which micrographia may occur, and its not infrequent connexion with disorders of vision and of the muscular sense.

In a paper ¹⁶ published in 1916, numerous instances were adduced of the association of micrographia or macrographia with the visual disturbance of dysmetropsia—the term proposed for departures from the normal either in the minus (micropsia) or the plus direction (macropsia)—and the occurrence of this visual defect at peripheral (lowest), cortical (middle), and transcortical (highest) levels was fully exemplified, cases of hysteria, migraine, and cerebral tumours, of tabes dorsalis and disseminated sclerosis, etc., being selected for the

God save the King

Have for and for ever

FIG. 13.—Micrographia in a case of moderate Parkinsonism.

purpose. With these clinical data before us, all that need now be said is that in cortical and transcortical conditions micrographia may be an accompaniment of macropsia, and, *mutatis mutandis*, macrographia of micropsia; or, alternatively, either variety of calligraphic defect may occur without visual disorder (as in the cases cited above). Again, micrographia often is met with where muscular rigidity is insignificant, as in some examples already given, and it may be seen in other instances where kinæsthetic factors have come into play. This latter association is of peculiar interest, for further details in regard to which the paper above referred to may be consulted.

To explain its admitted, if far from constant, occurrence in paralysis agitans and postencephalitic Parkinsonism, the following questions have to be answered: (1) Is it of transcortical, cortical, or subcortical origin, and (2) is it connected or unconnected with accommodation disorder? To take the latter first: Nearly all of

the Parkinsonians with micrographia that have come under notice have exhibited impairment of the convergence-accommodation mechanism, but none of them has complained of the visual disorder I call dysmetropsia. Besides, it is easy to show that in organic and non-psychical cases macrographia, not micrographia, is to be expected with micropsia, which is sometimes thought to result from paralysis of accommodation. For these reasons one cannot feasibly link the micrographia of Parkinsonism to any defect of convergence.

As for the former of the questions stated above, an analogy from this same visual field is apposite. I have shown that micropsia may result from lesions at one or other of three neural levels or, possibly, from lesions involving more than one. In a similar fashion we may discover the elements producing micrographia to have their anatomo-physiological site at one or other of the same three levels, or at more than one. Accordingly, we must take into account the possibility, as far as concerns Parkinsonian micrographia, of (1) infracortical disorders such as rigidity of antagonists; (2) central ("middle level") dysfunction, such as disturbance of pyramidal innervation for fine acquired movement; (3) "highest level," "transcortical" impairment, such as absence or diminution of the "will" to act, or the condition usually described as dyspraxia. Without further discussion in this place, enough has been said to prove that cortical and transcortical components may have as definite a share in the symptom under review as those of lower level origin, and that *acceptance of the striatal theory, in any exclusive sense, is impossible*.

(II) BEGINNING, COURSE, AND CESSATION OF MOVEMENT

That the Parkinsonian is "slow off the mark" is matter of common observation. Recent measurements¹⁷ with special apparatus for muscular response to a single visual stimulus have given the figures of 0.24 seconds for normal persons and 0.36 seconds for the subjects of paralysis agitans. With comparatively simple recording methods the feature of slowness in starting and continuing movement has been abundantly exemplified (cf. Figs. 5 and 7). In all the actions of daily life every Parkinsonian admits his unwonted delay in accomplishment. The "drag" is such as on occasion to bring volitional activity almost to a temporary standstill. Coupled with retardation is a restriction in range and excursion to which allusion has already been made.

In a large number of cases, however, this slowness is more apparent than real, and is so declared by the patient himself. The characteristic slow, shuffling, and limited gait can be modified in a surprising fashion if the observer puts his arm inside the other's and invites him to step out. The brachybasia may then easily disappear. A member of our profession, suffering from paralysis agitans, made the curious remark to me that he found he could walk more quickly over the stones of a rocky sea-beach than on the flat, the reason probably being that he was compelled to take longer steps. Parkinsonians often run better than they walk, in this respect presenting an illuminating contrast to tabetics, in whom inability to run while still able to walk is an early sign I have frequently observed. I have seen a subject of advancing paralysis agitans tuck up his feet and sprint along a garden path when progression at a walking pace was painfully slow. Another patient was actually able to proceed backwards, uphill, at a greater speed than forwards and on the level. James Parkinson gives a graphic description of more than one case in which "the inability for motion, except in a running pace, appeared to exist in an extraordinary degree." As a rule, this power of modification of an apparently hampered musculature is seen more in respect of the lower than of the upper extremities—a corollary, it may be, of the fact that the "voluntary" movements of the former are more, the latter less, "automatic." A remarkable circumstance connected with not a few cases of postencephalitic Parkinsonism has been variability in slowness and range-reduction at different periods of the day or night, at the instance of extrinsic or intrinsic stimuli, the action of which is not clearly understood. (In this connexion similar variability has been reported by one or two writers as taking place under the influence of hypnosis, but I am not in a position to add any personal contribution on this point. Improvement or disappearance of Parkinsonian micrographia under hypnosis has also been recorded.) In the idiopathic disease such peculiar variability is far less in evidence, but if we connect the changeableness in movement described above for the idiopathic patient with the curious though rare phenomena of the postencephalitic patient, we can only suppose the symptoms to depend less on structural lesions than on dynamic alterations of function, and that from time to time innervation is facilitated by modification of the background of muscular hypertonus against which it is ever contending. To these matters we shall return later. At any rate, we must concede the

possibility of fluctuations in the muscular state greater than a glance at the seemingly immobile attitudes might lead us to imagine.

As for cessation of voluntary movement, my observations lend no support to the view according to which the muscles in action in Parkinsonian cases may remain tonically contracted, perhaps for seconds, after the voluntary impulse has ceased. In no case have I been able to satisfy myself that any condition strictly analogous to tonic innervation occurs. By the latter expression is understood inability to decontract, to order or on a signal given, a voluntarily contracted muscle or group of muscles. Tonic innervation is of central origin, in the sense that the cases of it I have seen, and those reported by others, have been due to cerebral lesions, but as far as actual inability to relax a contracted muscular group is concerned, no clear clinical distinction can be drawn between it and the phenomenon usually termed myotonia. No criterion exists by which to judge when a contraction, voluntarily commenced, is prolonged sufficiently to be specified as "tonic," except that if it is maintained beyond the command to relax it is ipso facto pathological. Thus understood, tonic innervation in a mild form occurs in many morbid states (moderately severe hemiplegia, athetosis, etc.), but in my experience the phenomenon is so uncommon and uncertain in Parkinsonism as to be practically negligible. If the reader will look at Figs. 16 and 19, taken from cases of advanced bilateral postencephalitic Parkinsonism, he will find proof of this. In one case, that of a young woman, voluntary action involving the hands and fingers was feeble and tremulous and rather limited in range, but after sustained contraction of the long finger flexors ("close the fist") and, synergically, of the wrist extensors, at the word "open" the long finger extensors instantly contracted and the wrist extensors instantly relaxed—that is to say, neither protagonists (prime movers) nor synergists exhibited any trace of tonic innervation (Fig. 16). In this respect the behaviour of the muscles is identical with that seen in any normal subject (Fig. 15). Similar records have been obtained for the action of trunk and neck prime movers, as well as for trunk synergists in arm movements (cf. Figs. 18 and 19).

(III) POVERTY OF MOVEMENT

The term "poverty of movement" (*Bewegungsarmut*) has come into vogue to express the familiar symptoms of relative immobility

and stationariness of position which are particularly characteristic of subjects of the shaking palsy. No greater contrast can be imagined than is afforded by the akinesia of the latter, especially in the variety known as paralysis agitans sine agitatione, and the universal fidgetiness of the subjects of chorea. The Parkinsonian rarely if ever manifests spontaneity in change of position; he literally sits still. I have for a long time now been impressed by the fact that the average Parkinsonian never sits with his legs crossed, whereas nothing is more frequent in the normal person than a seated position with one knee over the other. Similarly, the Parkinsonian never sits with his arms folded, though here again is a commonplace change of attitude adopted frequently enough by the normal person. In a score of ways the absence of initiative, of spontaneity, of normal muscular play impresses itself on the observer. Among men a particularly common movement occurs in the course of ordinary conversation—viz. a movement of hand and fingers to chin, lips, and lower part of the face, but I have never yet seen a patient with paralysis agitans who has thus raised his hand. Women have a favourite spontaneous movement in similar circumstances—viz. toying with their wedding or other ring, but Parkinsonians do not do so. No changing expressions of emotion flit over the face—again providing a striking contrast with what obtains in chorea. This last remark raises an interesting point. Doubtless spontaneous movements contain “volitional” and “automatic” elements in inverse proportions—i.e. they are largely “automatic” and but faintly “voluntary”; on the other hand, change of position frequently results from the action of peripheral impressions on the sensorium, hence a conscious factor is not to be entirely excluded. But emotional expression is essentially involuntary, so that in the patient with Parkinson's disease we note the absence, or poverty, *both* of “voluntary” motor action *and* of “involuntary” motor action, at least as far as the expression of the emotions is concerned. I attach importance to this conclusion, and since a corresponding opposite is encountered in cases of chorea the point will be referred to again.

Allusion was made above to variability in the course of Parkinsonian movement from time to time, and a similar variability characterizes this “Bewegungsarmut.” On occasion, especially in postencephalitic cases, the patient appears to exhibit more freedom of movement than at others, and the ordinary spontaneous motions of everyday life are more in evidence. From patients of sufficient

intelligence and introspection it is possible to gain some insight into the causation of their motionlessness, for I may say at once I consider the subjective factors of no less importance and significance than the objective.

Causes of Parkinsonian Akinesis

(1) The general akinesis seems to me explicable largely by deprivation of, or serious reduction in, normal impulses to movement, both of the volitional and the spontaneous kind. When asked, patients will admit reduction of need for or impulse to muscular action. Their will to act is impaired. They say in so many words that movement requires a greater effort of the will than previously. Everything is done "with an effort." And the curious thing is the rarity with which they seem to wish to make such efforts; they appear to contemplate this defect of motor function with a philosophical resignation. Those afflicted with the disease complain more of losing the use of their limbs, of loss of muscular power and of capacity for executing the ordinary fine movements of every day, than of mere sitting still. We have seen that they can, as a fact, if they will, perform most of the movements of which superficially they appear incapable, but since this puts a tax on effort they simply do not make them.

Spontaneous change of posture being mainly a response to peripheral impressions, it may be designated a movement of reaction—a sort of psychoreflex movement, for consciousness is probably faintly stirred. To these sense-impressions the Parkinsonian and the senile (for a similar decrease in motility is natural to the senescent) are alike indifferent, probably for more than one reason. In the case of the former, no evidence suggesting an afferent origin for the impairment is forthcoming; the peripheral nervous system on the sensory side all agree is normal. A questionnaire addressed to a number of intelligent Parkinsonian patients has elicited the answer that they rarely feel the need to change position, and this might conceivably be taken to argue for an actual reduction in afferent stimuli, owing to relative fixity of the limb musculature. The muscles, however, are, many of them, in a condition of contraction; they are not resting or relaxed, immobile though the patient be; hence their sensory end-organs must be receiving stimulation. On the other hand, afferents from articular and periarticular tissues are probably to some extent restricted, but I cannot think this has a material bearing on the problem. That the immobility-effecting

disturbance of function is not sensory receives support from another consideration.

AKATHISIA

There are patients in whom immobility is actually a prominent symptom and who yet complain in a paradoxical way that "they cannot sit still," or only "with an effort." After a time they simply must get up and walk about, immobility having become intolerable. In other words, they feel, as does the normal subject, the cramping effect of unchanging posture and have to move and stretch their limbs at intervals; the accumulation or summation of afferent excitations, derived from a largely motionless (though not resting) musculature, is such that the patient must rise and "work it off," only, however, to sink once more into the same state of motionlessness. As, then, incoming impressions exist, reduction of spontaneity in movement must have its origin on the effector side. Since I commenced to observe this phenomenon in the course of my studies more particularly of the last two years, I find that Bing¹⁵ has directed attention to it, and employed the term originally devised by Haskovec—"akathisia"—for cases of inability to sit still of hysterical or psychasthenic origin. Bing refers to various authors who have noted akathisia in paralysis agitans and postencephalitic Parkinsonism, and while not committing himself to one explanation only, seems inclined to accept the view which some others have adopted and which is in accord with that here given. In my opinion akathisia is only an exaggeration of what is seen in normal individuals, and, since it has a psychogenic basis in some instances, it cannot be ascribed in any exclusive fashion to striatal disorder.

At this stage a word of caution must be entered as to indiscriminate use of the term "loss of movement" in connexion with Parkinson's disease. I have studiously avoided using the term "loss," apropos of reduction of voluntary and spontaneous movement, because I do not consider that the movements are in fact "lost" (with the few exceptions, and these only in part, already mentioned). *Because a patient does not make a movement it does not follow that he cannot.* It is of importance to bear this in mind when we discuss the functions of the corpus striatum at a later point.

(2) As for reduction of voluntary activity, the totality of sense-impressions at any given moment being in the Parkinsonian not much different from that of the normal person, we cannot ascribe the inaction to failure of extrinsic stimuli, although these doubtless

play a part as one of the springs of action. Intrinsic stimuli, at whose bidding the voluntary motor system passes into activity, may perhaps have an ultimate sensory origin, but we cannot express this impulse in physiological terms; we know only that our patients say they are not as conscious of it as formerly. It is idle, therefore, to speculate whether such intrinsic stimuli are damped down centrally by a process of inhibition which rarely lifts, or are not forthcoming in adequate amount, or not endowed with sufficient potential. Perhaps this potential has an affective quality, and the "affect-quantum" essential for an impulse to generate action may be wanting. Apart from considerations of this kind the Parkinsonian knows from experience that voluntary movements are no longer effortless and facile, and desists *largely of his own accord*. Even in respect of the sense of effort not a little is obscure. Binswanger, for example, declares that in paralysis agitans "innervation-feeling" is lacking, yet what is signified thereby is not clear. We have already noted the absence of sensory impairment in the disease, and cannot liken the motionlessness to that of the apæsthetic limb, all of whose dorsal spinal roots have been severed experimentally, as shown by Mott and Sherrington. Nor can sensations arising in the course of muscle innervation be lost; Parkinsonian muscles are not at rest. Once more, no evidence is available to indicate that motor or efferent currents are themselves sensed. "Innervation-feeling," therefore, or rather its absence, offers no explanation of the phenomena. I suspect that by a "sense of effort" the patient signifies little else than the consciousness that his muscles no longer "answer," as the rudder answers the helmsman's lightest touch at the wheel; and the conclusion, as indicated already more than once, is, that *some at least of the akinesia of the Parkinsonian is the direct outcome of unwillingness to move, since the making of movement is no longer effortless*.

(3) The objective state of the peripheral motor apparatus is without doubt a factor, yet only one, in this general akinesia. No constant relationship between degree of peripheral rigidity and decrease of spontaneous or of voluntary movement can be established, for the reason that the motor phenomena under discussion are frequently found where muscular rigidity is not a conspicuous feature. Relative immobility of facies and of limbs is often an early symptom in the disease, sometimes the earliest, before rigidity makes itself obvious. One of the earliest indications of the Parkinsonian mask is notable infrequency of ordinary blinking movements

of the eyelids, but in other respects facial movements, voluntary and emotional, may still be normal, no trace of rigidity in any facial group being detectable. Reference has not as yet been made to characteristic attitudes of limb segments in the disease, but it may be remarked here that they develop more or less *pari passu* with immobility—i.e., it is an immobility in attitude or posture. Now no constant relation obtains between rigidity and attitude in the first instance, for hands and fingers may assume attitudes involuntarily at an early stage when the corresponding muscles are not demonstrably rigid.

As the disease advances, however, a general tendency to muscular fixity becomes ever more obvious, implicating all skeletal groups more or less indifferently. It is due mainly to a hypertonic state of the musculature, an overaction of the tone-effecting apparatus, to which attention will be devoted subsequently. As a consequence, when the innervating current of "volition" reaches the motor periphery, it finds that: (1) The muscle elements it would innervate are already in relative contraction; (2) the antagonists to the required movement are also in activity. At this stage we shall not pursue the analysis further, for the matter can be more conveniently dealt with in the sections on disorder of muscle tonus. My observations, however, will show that in spite of the background of muscular overtone voluntary innervation can accomplish its ends as regards prime movers, synergists, and antagonists in a given movement, so that the conclusion is permissible that *the condition of the peripheral motor system is no more than one of the causes of Parkinsonian akinesia, and not necessarily the most important.*

We must beware in Parkinsonian cases of attributing to disease of the corpus striatum symptoms not in fact pathognomonic. A limb may be immobile, or relatively immobile, (a) because the lower motor neurones are paralysed, as in poliomyelitis; (b) because there exists a combination of innervation weakness and muscular spasticity, as in hemiplegia; (c) because normal innervation encounters a block in distal motor mechanisms, as in paralysis agitans (in part); (d) because central impulses are wanting, as in the same disease. Under (d) should be included, for the sake of completeness, certain cases of psychosis in which the immobility is psychogenic and partakes of a negativistic character. In short, *akinesia, or poverty of movement, obviously may arise at one or more of lower, middle, or highest physiological neural levels, a possibility which renders invalid its ascription to the corpus striatum alone.* Failure to en-

visage etiological differences in respect of the development of this symptom, as with others already investigated, has been a fruitful source of confusion ; with the exception of Binswanger,⁶ most recent continental writers have overemphasized the "hypokinetic-rigid" syndrome of the corpus striatum at the expense of other clinical conditions of which immobility is a symptom, but which are in no way associated with striatal disease.

(4) Somewhat similar explanations must apply to the general slowness, retardation, and limitation of voluntary movement in the disease, as well as to the muscular weakness. Here again a wide conception is indispensable. Allusion was made above to the "motor helplessness" of the subjects of paralysis agitans and of progressive lenticular degeneration, and in my original monograph on the latter disease the facts were fully exemplified. I found that "volitional movements may be good in range, but they are usually resisted with comparative ease. This muscular weakness is not to be confused with clumsiness or awkwardness from hypertonicity or tremor. There is intrinsic muscular asthenia ; the patient is incapable of any sustained effort ; he cannot close his eyes tightly for any length of time, cannot keep his tongue protruded, cannot maintain his grasp." At that time I endeavoured to avoid the use of the term "paralysis," hoping it might be confined to diseases of the pyramidal system. Further consideration has led me to modify this hope, for qua weakness, limitation, slowness, I see little material distinction between hemiplegic and Parkinsonian cases. The differences between pyramidal and extrapyramidal cases are distinct enough, but not as regards the above points. The *general* approximation of cases of double hemiplegia, especially of the pseudobulbar variety, to cases of paralysis agitans sine agitatione was emphasized long ago by Hughlings Jackson and again by Brissaud, and in these recent years we have seen examples of post-encephalitic Parkinsonism in which the diagnosis from pseudobulbar palsy has been more than a little difficult.

Having demonstrated the existence, in numerous instances, of such weakness, slowness, and imperfect range of volitional movement as constitutes, in Foerster's words, an "unmistakable paresis," I do not agree with him that there is no predilection type. On the contrary, a special class of acquired movements—viz., those of the smaller muscular groups throughout the body—is particularly liable to be affected, as has already been fully exemplified. The small muscles are those employed in what James Parkinson called

“ any nicer kind of manipulation ” ; their failure “ to answer with exactness to the dictates of the will ” is their characteristic in more than one state of motor defect. As a class, they suffer more than any other in hemiplegic cases ; derangement or outfall of fine finger movements is seen in hemiplegia, and of laryngeal, glossal, facial, and ocular movements (it may be) in double hemiplegia. In extrapyramidal, as in pyramidal cases, analogous but not necessarily identical imperfection of small movements is to be observed. It is true, as we have seen, that in some instances seemingly as definite paralysis of various small muscle activities characterizes Parkinson's disease, as it does pyramidal disease ; but no one considers the pyramidal morbid state the replica of the other. The rigidities are not identical, nor are the paralyses ; when, however, we find general resemblances in the types of movement that suffer, we are entitled to seek an explanation that is physiological rather than anatomical. In other words, *I do not take the weakness of certain movements in striatal affections to imply a concomitant anatomical defect of the corticospinal system, or of similar movements in hemiplegia to be the sequel merely of corticospinal interruption ; in each case the movements are disordered because their physiological quality or character is such that they suffer most and earliest.* To account for the weakness in paralysis agitans Foerster postulates a second corticomuscular path passing through the globus pallidus and in simultaneous action with the ordinary corticospinal tract in “ voluntary ” innervation. To me this hypothesis seems unnecessary (apart from its anatomical dubiousness) ; the rigidity in Parkinson's disease is *general*, but the weakness is *special*, and I submit the latter cannot be accounted for merely by anatomical interruption, just as in hemiplegia the special type of motor defect cannot be the outcome merely of a haphazard interference (by hæmorrhage or otherwise) of an anatomical system. In each instance there is defect or loss of *some* movements ; in each the same *sorts* of movement are disturbed ; behind both instances, therefore, must lie a similar disorder of function, but it is not a similar disorder of structure.

(For references, see list at end of Chapter XI.)

CHAPTER VIII

DISORDERS OF MOTILITY AND OF MUSCLE TONE, WITH SPECIAL REFERENCE TO THE CORPUS STRIATUM

II. *THE VOLUNTARY MOTOR SYSTEM IN STRIATAL DISEASE, continued.*

Behaviour of Muscular Components of Voluntary Movement in Striatal Disease. Movements of Sequence. Movements of Co-operation. Associated Movements. Arm-swinging in Walking. Presence of Certain Associated Movements in Striatal Disease. The "Blinking" Sign. Movements of Reaction and Defence. Movements of Expression. The Lore of the Antagonists, and their Rôle in Striatal Disease.

We now turn to a more precise examination of individual voluntary acts for differing parts of the body musculature, as an essential preliminary to an analysis of various kinds of involuntary movement. There is the more need for this investigation in that we are now approaching a clinical field where confusion reigns supreme in respect of nomenclature. Among the tangled mass of movements described by different observers as "primitive automatisms," "higher co-ordinations," "associated movements," "position alterations," "reaction reflexes," "reflex synergias," "movement synergias," "associated reactions," "automatized voluntary movements," "reaction movements," and what not, the student may well be excused if he loses his way.

Behaviour of Muscular Components of Voluntary Movement in Striatal Disease

I have already indicated the lines I shall follow in this connexion. I shall take a normal movement to have as constituents the actions of prime movers or protagonists, synergists (including fixation muscles), and antagonists respectively. I shall not call the whole a "synergic unit," or a "movement synergia," but simply a movement, with the component parts just enumerated.

The following are the single or combined movements studied in normal persons and in a large number of cases of striatal disease.

I. Head, face, and neck.

1. Shut eyes, with or without resistance; eyeballs roll up.
2. Turn eyes to side; head turns also.
3. Open mouth, against resistance; head extends.
4. Turn eyes up; forehead wrinkles, head extends.
5. In supine position, flex head on chest; recti abdominis contract.
6. Flex head against resistance; platysmas, omohyoids, hyoid depressors contract.
7. With head in one extreme lateral position (say, right) rotate it slowly to left; left posterior rotators contract first, and right sternomastoid only after head is well past the mid-line. And vice versa.

II. Shoulder, arm, hand, and fingers.

1. Abduct extended arm at shoulder; opposite erector spinæ contracts.
2. Adduct extended arm from horizontal position; homolateral rectus abdominis and erector spinæ contract (only if against resistance).
3. In erect position, advance extended arm to horizontal level in front—both erectores spinæ contract; rotate it horizontally backwards—homolateral erector spinæ at once relaxes; bring it forward again on horizontal level—the same erector spinæ contracts again; depress extended arm once more to side—homolateral erector spinæ once more relaxes.
4. With arm flexed at elbow and forearm pronated, supinate (biceps does this); triceps contracts to prevent further flexion at elbow.
5. Flex fingers in grasping; extensors of wrist contract. Increase grasping movement; triceps contracts.
6. Open closed hand; extensors of wrist relax: flexors of wrist contract.
7. Extend flexed wrist with fingers closed—extensores carpi radiales and ulnaris contract; without stopping wrist extension, open fingers—the wrist extensors at once relax, while the extensors of the fingers continue the upward movement.
8. Press thumb firmly against forefinger; flexor carpi ulnaris contracts.
9. Abduct little finger; extensor ossis metacarpi pollicis contracts.
10. Extend thumb; extensor and flexor carpi ulnaris contract.
11. Abduct forefinger; thumb nearly always flexes or adducts and, if so, extensors of thumb contract.

III. Trunk and legs.

1. Supine position; lift one extended leg (i.e., flex at hip); contralateral hamstrings (internal or external group, or both) contract.
2. Supine position; lift head and shoulders up; recti abdominis contract, then both recti femoris.
3. Erect position; abduct one extended leg at hip; opposite tensor fasciæ femoris contracts.

4. Flex spine—i.e., bend forwards (not against resistance); both erectores spinæ contract.
5. Bend backwards—i.e., extend spine; recti abdominis contract. Combining (4) and (5), recti and erectores alternately contract and relax.
6. Incline spine to right or left; opposite erector spinæ contracts.

This scheme contains a description of the precise behaviour of muscular components in a number of normal movements and movement-combinations, and has been carried out in extenso in many examples both of paralysis agitans and of postencephalitic Parkinsonism.

With minor qualifications to be referred to immediately, my definite conclusion is that *these movements are executed normally by Parkinsonians—that is to say, the movements follow normal physiological lines and are neither lost nor perverted*. I have not seen an instance either of failure to perform the movement (except as below), or of interference with normal physiological order. In what might be termed *movements of sequence* (viz. I. 7, II. 7, III. 5 above), I have not found any departure from the normal. By this term I mean movements begun by one muscle or muscular group and continued by another. My former chief, the late Dr. C. E. Beevor,¹⁰ devoted attention to these, showing the order of action to be so fixed physiologically that it cannot be voluntarily modified, and that if one muscle in the total act fails its place cannot readily be taken by another which might appear suitable for the purpose. He said, “it rather reminds one of some highly organized establishments where if one of the servants is incapacitated it is no one’s place to do the work, which is consequently left undone.” The qualifications I would make are, that if the action of agonists is inherently weak that of the other components is correspondingly feeble and therefore difficult to recognize, though as a general rule the introduction of an element of resistance against which to work intensifies the voluntary effort and so may bring out the constituents better. For example, I find the triceps often will not partake in the hand-grasping movement (II. 5 above) because the latter cannot be executed with sufficient power. Further, some finger movements, notably abduction of the little finger, may in fact almost fail, for reasons already sufficiently gone into. Again, in some normal persons part of the movement is facultative rather than obligatory—e.g., in I. 4 the forehead may not wrinkle; we must avoid, therefore, an over-estimation of such “loss” in Parkinsonian cases. The study of these movements, finally, in seriously advanced cases is open to the

objection that myogenic contractures may have set in, and this will naturally vitiate the clinical findings.

The main conclusion stands, and has weight. As a consequence, *the theory* (Foerster and others) *according to which* "loss" of these "*Mitbewegungen*" (an unfortunate terminology), *in reality the components in a total single movement, is held to prove that the corpus striatum is a centre for their normal activation, is shown to be based on erroneous observation and to have no foundation in fact.*

I may take this chance of again entering a caveat against the view that because muscular action is hindered or encounters obstacles it is "lost." In some instances of excessive muscular rigidity, especially among postencephalitic cases, the movement of the head accompanying lateral deviation of the eyes is minimal, yet to describe it as lost and at the same time to ignore the multiplicity of factors in Parkinsonian immobility would be a mistake. Of the comparative slowness with which the above enumerated movements are sometimes performed there can of course be no question, nor of the frequent weakness of some of them. In respect of slowness, records from unilateral cases of postencephalitic Parkinsonism are instructive. In the movement of raising the shoulders from the supine position (III. 2 above) the normal subject contracts both recti femoris simultaneously and within a very brief period after shortening of the recti abdominis. In unilateral cases delay in contraction of the rectus femoris on the affected side has been constantly found, but I have failed to note any difference in this respect in that of the twin recti abdominis. Fig. 14 gives a tracing from a case of this kind, showing on the Parkinsonian side a delay of about three-quarters of a second in the contraction of the muscle on the front of the left thigh compared with that on the right (unaffected) side. This difference in the case of the two recti femoris is striking, though they contract with equal *power*, as the curves indicate.

In some instances the reverse obtains; weakness is discernible while slowness of contraction or relaxation cannot be detected. We have seen (II. 6 above) that when the closed fist is opened wrist extensors immediately relax while finger extensors contract. Fig. 15 shows the constituent actions as they occur normally, relaxation of the former and contraction of the latter being simultaneous. Figs. 16 and 17 are from cases of Parkinson's disease; in Fig. 16, on the signal given, wrist extensors are seen to relax at the same moment as finger extensors contract, without retardation, and notwithstanding the tremor with which the movement is

accompanied. In Fig. 17 the lower tracing shows gradual weakening of contraction of wrist extensors before the signal—in other words, the *inability to maintain an innervation* which often characterizes Parkinsonism. On the signal, however, the normal procedure at once occurs, though decontraction of the wrist extensors is

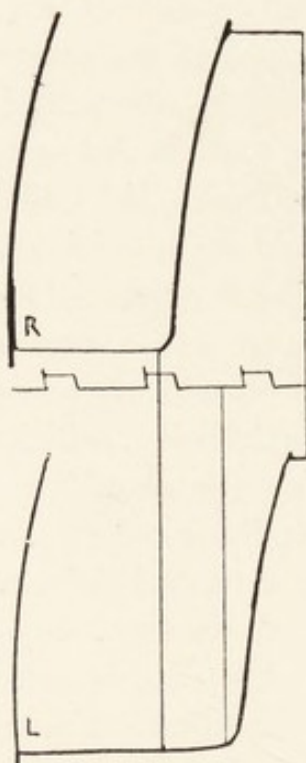


FIG. 14.—Curves of contraction of the recti femoris in the combined movement of sitting up from a supine position. Unilateral Parkinsonian case.

The affected muscle (left) contracts about three-quarters of a second later than that on the normal side.

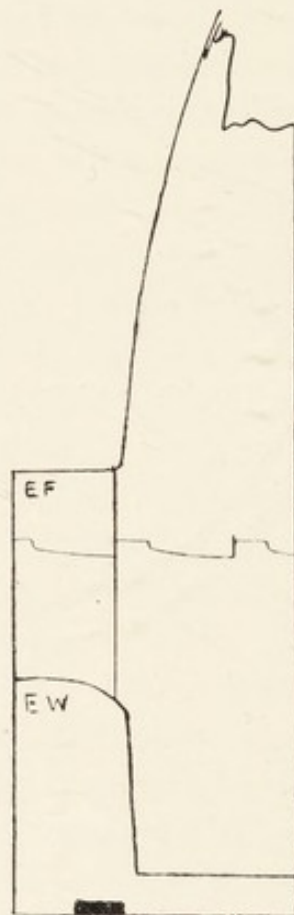


FIG. 15.—Normal hand, with fist closed voluntarily.

On the signal ("open the hand") the extensors of the fingers (EF) contract and the extensors of the wrist (EW) relax simultaneously.

fractionally slower than contraction of the finger extensors and exhibits the so-called "cogwheel" phenomenon (to be afterwards discussed).

Equally instructive are tracings of the combined movement described under II. 3 above. The extended arm (say, left) is advanced to the shoulder level in front, swept back in an arc on this horizontal level, brought forward again, then down to the side. As this is being done the left erector spinæ contracts, relaxes, contracts

again, relaxes again. Fig. 18, from a normal subject, is to be compared with Fig. 19, from a severe case of chronic postencephalitic Parkinson syndrome. It must be borne in mind, to understand the meaning of the tracings, that in the erect position the erector spinae of the normal person are not in action; in the Parkinsonian,

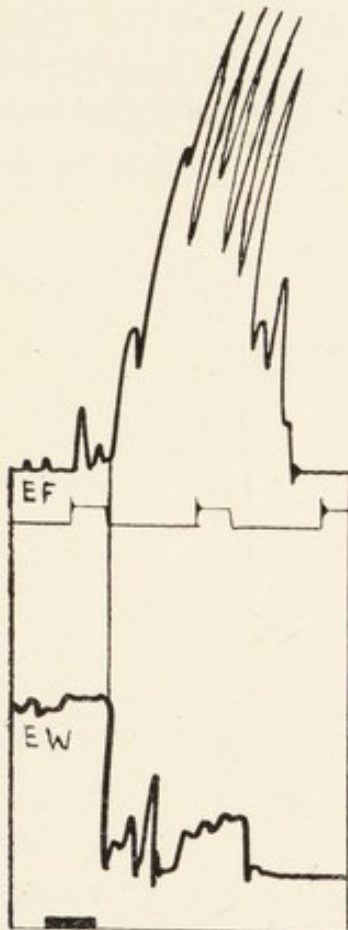


FIG. 16.—Case of severe Parkinson's disease.

On the signal ("open the hand") the extensors of the fingers contract and those of the wrist relax simultaneously, in spite of rigidity and tremor.

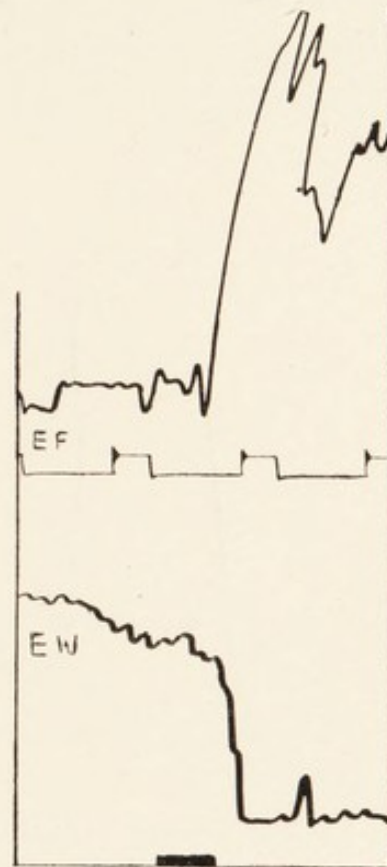


FIG. 17.—Similar case to Fig. 16. Same correct physiological interrelation of extensors of fingers and of wrist.

Note how the voluntary contraction (synergic) of the extensors of the wrist weakens before the signal (inability to maintain a voluntary contraction).

with his typical forward inclination of the trunk, they are, on the contrary, in strong (involuntary) contraction. Now when the two records are examined the reader will see that, notwithstanding steady postural contraction of the spinal muscle in the Parkinsonian, its successive voluntary contraction and relaxation follow normal physiological lines; it rises and falls exactly as does the normal erector spinae, at the appropriate signals. Particularly instructive are the two points (1) that at the beginning of the "voluntary"

movement the already posturally contracted erector spinæ *contracts still further*; (2) that when it is beginning to relax normally as the movement is coming to a close it springs again into this steady "involuntary" contraction. (Although the Parkinsonian record is slower than the other the two are not in this respect comparable, since no attempt was made to have the patient perform the movement quickly.) The actions of the erector spinæ during execution of the combined movement are not characterized either by undue retardation or by limitation of range; that is to say, that component of a "voluntary" movement of whose synergic action the patient is



FIG. 18.—Normal subject. Combined movement No. II. 3. Tracing of contractions of homolateral erector spinæ.

(a) Extended arm advanced to horizontal level in front: e.s. contracts; (b) arm swept back on same level: e.s. relaxes; (c) arm brought forward again: e.s. contracts; (d) arm brought down to side: e.s. relaxes.

unaware (no one in thus advancing and rotating the arm has any idea that the erector spinæ constantly partakes therein), and which, as indicated at the outset, is "involuntary" in the sense that it is invariable and outside consciousness, can and does impress itself on the existing background of postural contraction, modifying the latter in the course of its own appropriate action. A single instance of this description shows incidentally the unsatisfactory character of the usual distinctions drawn between "voluntary" and "involuntary," for we have here the idea of *degrees of involuntariness*, one major and the other minor, one part of a so-called voluntary act, the other part of a so-called postural contraction. Further, these illustrations serve to lend point to the statement made in the intro-

duction, that in objective characters, clinically speaking, voluntary movements are not to be distinguished from those in which the element of volition is lacking. Neither the examiner's finger nor the myographic tracing reveals any difference between the contraction of the erector spinæ at (c) (Fig. 19), and at (d) (same Fig.), but the former, according to the usual conceptions, is "voluntary" and the latter "involuntary." As a clinician, I cannot say where volitional activity ends and non-volitional activity begins.

The conclusion is, that *notwithstanding weakness, slowness, rigidity, and tremor, the ordinary muscular components of voluntary*

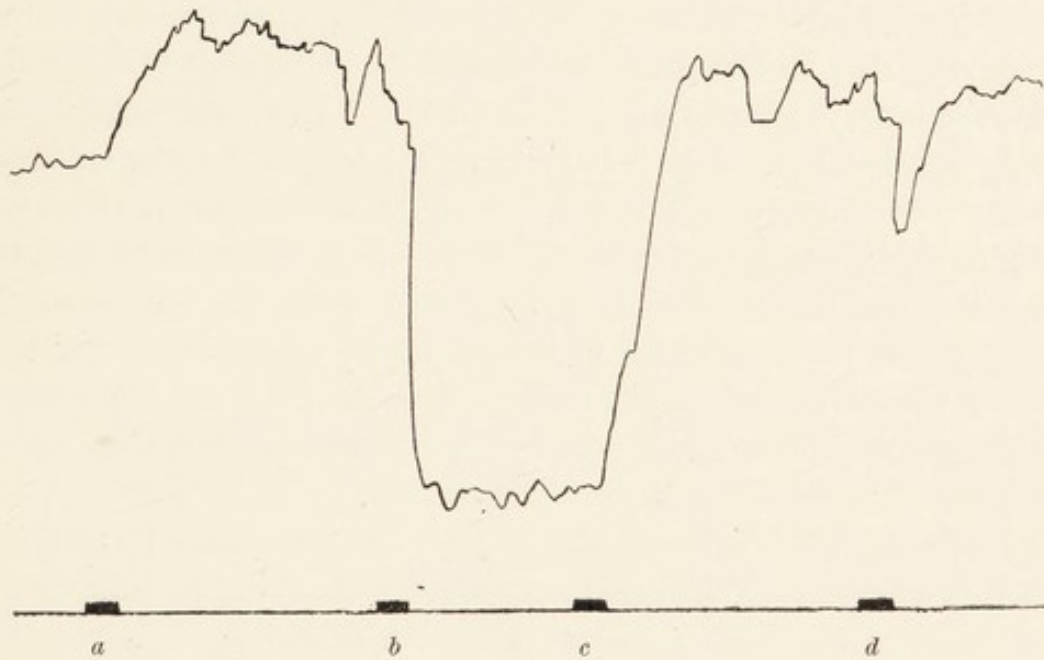


FIG. 19.—Parkinsonian case. Combined movement No. II. 3. Compare with Fig. 18. (For details, see text.)

movement behave normally in cases of striatal disease, contrary to the views of not a few writers on the subject.

We may now pass to the consideration of another class of movement, which I shall call

Movements of Co-operation

By this expression I wish to signify movements that are auxiliary to other, also voluntary movements, preceding or accompanying these as the case may be. Thus when a person seated on a chair is requested to stand up he first of all brings his feet in underneath his body, to a sufficient extent to place them under, or almost under, his centre of gravity. An essential preliminary to the main movement, the rising up, it aids the latter by enabling it to be performed

with appropriate ease and may therefore be described as a movement of co-operation. Other writers have called it an "associated movement," an "automatic movement," a "reaction movement," and have declared specifically that such are defective or lost in disease of the corpus striatum. But none of these alternative expressions will stand criticism. No one can say when a movement passes out of the voluntary into the automatic group. If I have to go down an unknown staircase in relative darkness, I will gauge the height of the first few steps by feeling with my feet; having estimated it I will probably thereafter proceed more quickly, but I do not pass from a voluntary to an automatic movement thereby, as if these are two different *sorts* of movement; my stepping movement, a voluntary movement, is "variably automatic." "Voluntary," applied to movements, means nothing else than "least automatic," as already pointed out. Nor is it appropriate to call the movement of drawing the feet under the body a "reaction movement," as Foerster does; in one sense all movements are reaction movements, and the expression becomes meaningless. I shall reserve the term for another group (to be referred to immediately) where definite stimuli are used to provoke response. Finally, as we shall see, the expression "associated movement" is equally devoid of specificity unless it is properly restricted in its connotation.

This movement of co-operation, then, I have found to be as a fact present in all cases of Parkinsonism provided the feet are, prior to the assumption of the erect position, placed a little way out from the chair. Applying the test to normal persons, I have noticed that they can rise up without this auxiliary and preliminary leg flexion as long as the knee angle does not exceed, say, 135° ; even then, only with an obvious effort do they "throw themselves" up. Exactly the same principle applies to the Parkinsonian; since he rarely sits with limbs extended, his customary attitude being one of flexion, his feet are already in a suitable position as a rule, and he can rise without the auxiliary flexion, but put them out a little and invariably he brings them in to begin with, slowly and unequally as may be. *To say that this and analogous movements are lost and to argue therefrom that the corpus striatum is a centre for "automatic" or "associated" movements is, therefore, a fundamental mistake.* Similar remarks apply to the slight outward movement of the feet (extension of the legs) taking place almost invariably after the normal person has resumed his seat, provided his limbs are close to the chair as he sits down. (This movement, however, is scarcely

one of co-operation, but rather of relaxation, since sitting with limbs flexed at knees to less than a right angle may become uncomfortable.) The common flexion attitude of the Parkinsonian explains the usual absence of the little outward extension, but in not a few cases (and these not instances of only moderate rigidity) it is readily observable. In one case of bilateral Parkinsonism (left side more affected than right) the movement constantly occurred on the left side, but never on the other.

Equally interesting is the swinging action of the arms in walking, and in a way more important, in view of the fact that it is constantly referred to as an "associated movement," and its supposed loss in striatal disease as constantly taken to argue the existence of a physiological centre for "associated movements" in the normal corpus striatum.

Associated Movements

Few terms are as much in vogue and at the same time as vague in meaning as this is, and its inherent indefiniteness will remain, unless, indeed, each writer will specify the exact sense in which he employs it. In ordinary neurological parlance the term, I take it, is used to denote those involuntary movements of one limb, say one arm or segment of the arm, which may be seen to accompany forceful voluntary movements of its fellow on the other side. Such movements have long been known to be readily elicitable, more particularly in cases of hemiplegia and diplegia—in the former condition usually from the *sound* to the *affected* side, in the latter indifferently from either. The associated movement on the second side may, or may not, be a mirror movement of that on the first side; it often spreads on the second side in a way not to be seen on the first. Foerster speaks of them as "the normal [sic] synergias of pyramidal cases"—an unfortunate expression, in view of the fact that they are found in diseased states. It is confusing to find included by Foerster³ under the same term those muscular actions (extension of the wrist during closure of the fingers, etc.) which we have seen are precisely the normal muscular components of a total single normal movement. Moreover, this author places in the same category of "normal synergias" the spread of movement in hemiplegic cases from one group to another in the *same* limb, as when such a patient cannot flex his forearm without pronating and abducting at the same time, and also the "mass movements" of tetraplegia and spastic paraplegia. No distinction, apparently, is

drawn by him between the "associated movements," in his sense, of normal persons and of hemiplegics, normal and abnormal being classed together; and, finding them both wanting, as he considers, in pallidal cases, he reaches the conclusion already indicated, that the normal corpus striatum is the centre for this type (these types) of movement.

There is here, I consider, a plain error of deduction. Most observers are agreed on the general absence in striatal cases of the above-mentioned and well-known associated movements of pyramidal cases—widespreading, tonic, reflex movements which are the result of release of function of lower and older motor mechanisms, and which are abnormal in the sense that for their appearance a pathological condition of the corticospinal system is usual. But it is illegitimate to maintain that the absence in *striatal* disease of pathological associated movements proper to the diseased *pyramidal* system argues for their localization in the basal ganglia. Taking Foerster's own statement, that they are also to be found in the form of "mass movements," we know the latter may be seen in cases of total transverse lesion of the cord—a condition with which the corpus striatum has nothing to do in any direct sense, since it is anatomically severed and as much out of action as in destructive striatal lesions. The conclusion is unmistakable, that *this type of "associated movement" is not effected through the globus pallidus and has no connexion therewith.*

As for the ordinary "movement synergias" of normal persons—i.e., the normal muscular components of a single movement—the "normal involuntary associated movements of agonistic synergists," in Foerster's elaborate phrase—we have already found them to be typically present in Parkinsonian and other striatal cases, and it is therefore impossible to subscribe to his theory, that they are effected by sensory impulses streaming to the globus pallidus via the optic thalamus from the beginning to the end of voluntary movement and ipso facto disappear in disease of that part of the basal ganglia. This theory is based on what I consider to be erroneous observation.

ARM-SWINGING IN WALKING

Leaving pathological (pyramidal) associated movements and normal components of voluntary movement (which ought never to be classed in the group under discussion) on one side, we next ask ourselves whether any other variety remains. And here we may revert to the question of swinging movements of the arms in

walking. My own investigations lead me to suggest they should be regarded rather as *movements of co-operation* in the sense defined above. They are auxiliary movements, practised to enable the individual to maintain balance better during the act. Though normal, they are in no respect invariable, for the simple reason that anyone can walk with his hands clasped behind his back, or with his arms held stiffly in front or in any other artificial fashion. He will at once, however, be conscious of lack of the ease in walking thus, and will revert with relief to normal swinging. At the other extreme is the powerful use made of the swinging arms by expert athletes in walking races, when the upper limbs can be seen to be voluntarily utilized by exaggerated sweeping movements across the chest with a view to facilitating progress. I cannot assent, therefore, to the opinion either that this arm movement is a constant normal "associated movement," or that it is purely "automatic."

Of its common reduction or cessation in Parkinsonian cases let me state at once there can be no question. Normally the movement, I find, largely consists of moderate flexion at the elbow, with but slight advance of the limb at the shoulder. On the whole, the arm moves through a wider range in front of the mid-axillary line than behind it. Now if a Parkinsonian be watched as he walks with body stripped, absence of any forward movement at the shoulder will at once claim the observer's attention, and as a slight advance at the proximal end of the limb would of course result in a materially wider excursion at the distal end, its failure to occur will affect the whole limb. In a fair number of instances, however, some flexion at the elbow takes place. In other, asymmetrical, examples of the disease the less affected arm will move with some freedom as compared with its fellow; in unilateral cases the sound arm will move normally enough, but this class of case sometimes presents unusual and interesting features. A moment's thought will remind us that the normal arm is moving with the impeded leg, and I find on observation of several such examples that movement of the sound upper limb is made to correspond with and compensate for defect of the opposite lower limb. Thus in one case (largely right-sided) the left arm was always advanced to a distinctly *greater* extent than usual, and this corresponded strictly to a slower advancement of the weak right leg. In another case (also right-sided), the normal left arm was never quite at the side, but was held in advance continuously and made a swinging movement

through a rather reduced range in this advanced position. In unilateral instances movement of the sound arm is "variably automatic."

It is instructive to note how frequently intelligent patients are aware of this imperfection of arm-swinging during their walk, even at a comparatively early stage, but if they are asked deliberately to swing the arms—"as they should do"—curious effects may result. For example, while some will manage for a time to move the arms alternately and appropriately, the movement will have a somewhat unnatural look; others, again, as I have seen more than once, move the arms forward and backward together in a ridiculous way; another patient (unilateral left-sided case), told to swing the left arm "properly" as she walked, did so only at the expense of the right arm, saying, "if I try to swing my left arm my right arm stops swinging." Patients, too, who when given a similar request hold both arms forward with a minimum of swing have been observed. In one case the patient made the spontaneous remark that the left arm (affected side) "felt different"; "in walking I feel as if the left arm has no weight, as if it were hanging by a thread." Another made the candid and illuminating observation: "It is easier not to swing my arms."

The explanation of this frequent though not universal phenomenon is to be found by reference to more than one consideration. Undoubtedly the factor of proximal rigidity plays a significant rôle, as is immediately evident when the patient walks stripped; involuntary contraction of deltoid and upper arm muscles keeps them relatively tense, and no alternating "play" between anterior and posterior groups is observable. I have not yet seen a case of reduction of this specific arm movement in which these muscles could be said to exhibit normal muscle tone. The hemiplegic patient does not swing his rigid arm. The matter of muscular effort, again, cannot be ignored, and the same remarks are applicable in this respect as have been offered above, apropos of the Parkinsonian's general immobility. Largely automatic though the arm-swinging be, it encounters resistance; hence not to do it is "easier." In forcing the movement deliberately the Parkinsonian encounters the same difficulty as with any of his voluntary movements, and the result is one or other of the various anomalies already enumerated. Nor must it be forgotten, as before-mentioned, that arm movements are not essential in the walking act, and since the patient can as a fact walk without their aid the faint element

of conscious volition which should assist in stimulating them to action is ignored.

My conclusion, therefore, is to the effect that, *this arm-swinging being neither constant, essential, nor purely automatic, and partaking more of the nature of movements of co-operation or auxiliary movements than of involuntary or reflex associated movements, its reduction or absence in cases of striatal disease cannot be regarded as in any way supporting a theory of the localization of "associated movements" or of "automatisms" in the corpus striatum.*

Presence of Certain Associated Movements in Striatal Disease

Finally, under this heading, I wish to direct attention to the frequent or almost constant occurrence, mainly in unilateral cases of Parkinsonism and allied conditions, of a variety of associated movement from the *affected* to the *sound* side, but not, as a rule, vice versa. Many times have I had occasion to observe how in the performance of individual movements with the affected arm, hand, or fingers (especially in the case of the latter two) an identical mirror movement is executed on the sound side, smaller in range as a rule and usually at least as slow as the other—sometimes, in fact, more prolonged. For instance, abduction of the little finger on the weak side is accompanied by the same movement of the normal finger, extension of the wrist on the affected side by extension of the wrist on the other, and so on. In ordinary bilateral cases these phenomena are often wanting, but should one side be more affected than the other they are commonly still to be seen, always from the more weak to the less weak side. So much is this the case that the observer can thus sometimes decide, by a rapid and simple test, which of the two sides is the more involved. The tracing reproduced in Fig. 20, taken from a unilateral case, shows how flexion of the forefinger of the affected arm (below), relatively good if somewhat tremulous, is accompanied by a fine flexion of the forefinger of the normal hand, beginning a fraction of a second later, considerably more restricted in range, and, in this instance, continuing a second or two after the movement of the first side has ceased. (Cinematograph films that I have had taken give more striking examples of the crossed muscular action.)

Movements of this character have been described in another connexion by Pierre Marie and Foix¹⁸ as "*syncinésies d'imitation*," and are stated to occur indifferently from either side in cases

of hemichorea and hemiathetosis. The peculiarity here remarked is their occurrence mainly *from the affected*, or more affected, to the *sound*, or less affected, side in cases of striatal disease, which, according to the usual views, are characterized by absence of all associated movements. Comment has already been made on the loose way in which this expression is employed, so much so that it is in danger of becoming valueless; but while the phenomena just described differ in some respects from the familiar involuntary associated movements of pyramidal affections they clearly exhibit the element of association and deserve attention. They can be, and often are, inhibited, especially if the subject becomes aware of the examiner's attention to the crossed movement rather than to

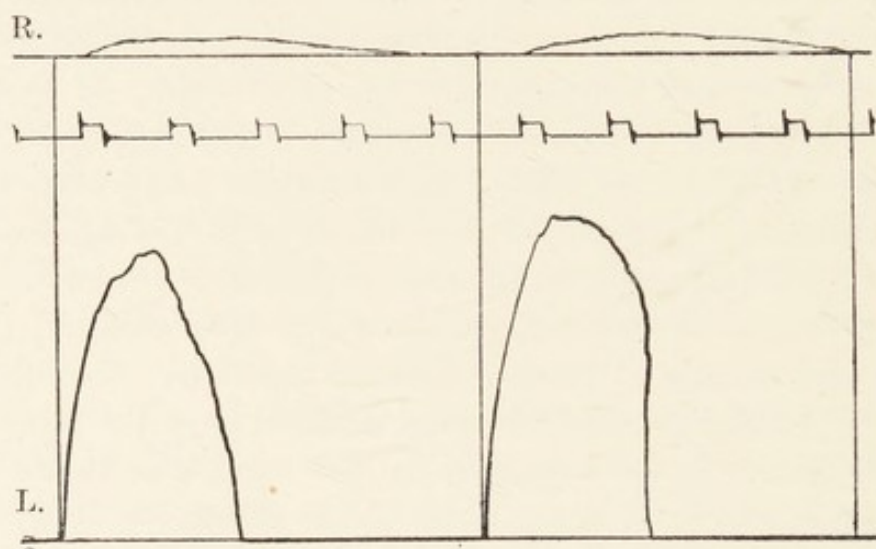


FIG. 20.—Unilateral Parkinsonian case. Associated movement of right (normal) index finger during voluntary flexion of left forefinger (affected side). The associated contraction begins later, is much less in degree, but lasts longer.

the requested movement of the weak limb; but they are not found to occur in normal people, where individualization of movement is perfectly developed, and I do not believe, as do Marie and Foix, in the existence of a "normal tendency" to imitation by one side of the movements of the other. Simple though the movement is, a satisfactory explanation is not easily found. The conjecture might be hazarded that the peculiar unilaterality, the occurrence mainly on the unaffected, or less affected, side, points to an overflow of innervation at a low (cord) level to the corresponding motor centre of the other side, conceivably because of the relative block encountered by it in the (affected) muscle units to which it is directed; this tendency to cross-innervation is readily inhibitable from the cortex. Since no such obstacle is met on the healthy side, there is

no tendency when *it* is innervated for the current to cross to the affected side. I do not think that any explanation based on bilateral representation of individual movements in the cortical motor centres applies here; we cannot say it is in any way customary for such corresponding muscular groups to be innervated simultaneously and identically, at least in the case of the muscles of fingers and hands, where marked individualization is the rule.

THE "BLINKING" SIGN

In further contradiction of the general opinion as to loss of associated movements in striatal affections, a word may be said on a peculiar variety of synkinesis which I first noticed a number of years ago in cases of paralysis agitans and which has been repeatedly observed in patients suffering from the diseases of this group, while it is according to my investigations of but rare occurrence in normal persons. When the Parkinsonian is asked to make a series of successive right and left conjugate deviating movements of the eyeballs (the head being kept steady) he very frequently blinks his eyelids as the eyes pass from one extreme lateral position to the other. The blinking movement, of which he appears to be unaware, is sometimes quick, sometimes slow and deliberate. In more than one case a quick double eyelid flicker has been distinctly and repeatedly observed. It is less commonly noted on upward and downward movement. This transient complete or incomplete eyelid closure is the more noteworthy in that Parkinsonians as a rule are conspicuous for the poverty of blinking movements, as has been made the subject of previous allusion. Now normal subjects, who blink easily and readily, do not show this particular associated movement to anything like the same extent, though I do not say it is never seen. So high a proportion of Parkinsonian patients exhibit it that I have come to regard it as more typical of that condition than of any other. Yet no explanation of this eye-blinking associated movement that I can think of can be said to be satisfactory, and for the present I am content to note its existence.*

Summing up our clinical data and our deductions therefrom, we

* A young woman, recently seen, suffering from postencephalitic Parkinsonism, exhibits extremely slow, incomplete, and punctuate lateral deviation of eyes to right and left, but she has discovered for herself that if she lets her eyelids droop, or shuts the eyes, as she deviates the eyeballs conjugately from side to side, their movement becomes at once quicker and more complete.

are justified in the general conclusion, that *striatal disease is not characterized by complete outfall of associated movements ; we naturally do not expect the special variety discoverable in pyramidal affections to be present in extrapyramidal disease and therefore cannot in any sense describe it as lost ; other varieties are actually found in striatal maladies and not in normal subjects ; others still have been erroneously included in this class of movement and any argument based on their reduction or absence is thereby vitiated.*

Movements of Reaction and Defence

In this category are comprehended movements in response to definite stimuli and those performed by way of protection or defence from some nocuous excitation. Such motor responses are frequently designated "reflex movements," but this term is undesirable. In the intact organism an immediate response to a nocuous stimulation is called reflex, as when the eyes are closed on the flash of a bright light (in any flashlight group photograph a percentage of the sitters are seen to have their eyes shut). Yet in numerous instances a conscious element cannot be said to be excluded. The hand is moved with urgency from the contact of a hot surface, but the sensory impulse must reach the cortex first, there to awaken a conscious sensation of heat. Otherwise expressed, the reflex arcs extend to the cortical level, and if so it is perhaps preferable they should be distinguished as a class from reflexes confined to lower physiological levels, however analogous their mechanism.

The general statement is frequently made by those concerned to defend current conceptions of striatal function that in striatal disease all movements of reaction and of defence ("automatisms") are impaired or lost, whence we may infer the tacit assumption is drawn either that consciousness accompanies striatal activity or that striatal "automatisms" blend in some undefined way with higher (cortical) co-ordinated movements. My own observations in this respect also lend no countenance to the theory of striatal function erected thereon.

When testing for the class of movements now under review it is imperative the subject should have no cognizance of what is about to be tried out. I devised a series of chair-tilting tests in which the patient's chair was abruptly moved in one direction or another by an assistant behind him, while the observer was seated in front. The degree to which conscious knowledge of the tests to be made interferes with the reactions is sufficiently indicated by the remark

of a patient whose responses became less and less as the examination proceeded and who, when comment was made thereon, said, "I know now you won't really let me fall off." Cases of severe generalized Parkinsonism were selected for the purpose, the patient being seated on a chair with hands on lap and then tilted abruptly forwards or backwards or to one or other side. A single series may be briefly summarized to illustrate results. In this instance the case was that of a woman of 28, suffering from the postencephalitic variety.

(1) Tilted to L. The left leg was at once abducted, the left arm extended, abducted away from the trunk, and directed towards the corner of a table which stood near. As further falling to the left continued, the right arm was in turn brought across and directed also to the table. There was no movement of the right leg.

(2) Tilted to R. The right arm was immediately extended and abducted and the fingers extended as if to save from the floor. The left leg was notably abducted, extended, and elevated, as if to counterbalance the right lateral deviation. No movement was observed in left arm or right leg.

(3) Tilted backwards. Both arms were at once thrown out in extension in front, both legs were abducted, and slightly extended at the knees.

Pushing tests were utilized in the same case, the subject having the eyes closed to obviate foreknowledge.

(1) Sudden push backwards. Both arms were thrown out in front in full extension.

(2) Sudden push to L. and R. respectively. In each instance there was marked quick abduction of the ipsilateral leg, with slight abduction of the contralateral arm.

All sorts of variations have been seen in different subjects, and in some the reactions have been considerably restricted as compared with the above. Thus in the case of a man of 27 (post-encephalitic form) in whom the left side was distinctly more affected than the right, chair-tilting to the *left* evoked abduction of the *right* leg, then feeble abduction of the left, while the edge of the chair was gripped with the right hand; whereas, on tilting to the *right* only the *right* leg was abducted, and no movement of the left limbs was detected. On sudden pulsion backwards the same individual's reaction was confined to slight but definite advancing of the right arm. Another patient whose responses to these tests seemed peculiarly meagre (a girl of quick intelligence, she early realized that she would not be allowed to hurt herself), was asked to bend

backwards as far as she could, on the pretence of examination of the back muscles. The supporting hands were suddenly removed, when at once both her arms were thrown forward and she tried to catch hold of the assistant standing in front.

In view of these findings, which might be readily amplified, we shall be careful in our interpretation of the seeming absence of reaction among Parkinsonians to peripheral stimuli of differing sorts. Foerster, for example, says they exhibit no movements of reaction to irritation from flies; but neither do the moribund, nor the profoundly asthenic from serious illness, nor those in whose case for other physical reasons the reaction requires an effort. "It is easier not to." Indifference and apathy led to similar absence of reaction on the part of the long-suffering soldier on the Gallipoli Peninsula. In short, *so manifold are the factors leading to this apparent poverty of reaction movements, and so clearly is there resident in them a cortical, because conscious, element, that their ascription to striatal function can be dismissed as a conjecture lacking inherent probability no less than any clinical support.*

The question can be further probed by resort to the familiar tests for defence-reactions to stimuli along the sense avenues of touch, sight, hearing, etc. With the proviso that the subject must be unaware of what is to be examined for, the nostril, ear, or throat can be tickled, visual or auditory excitations of a sufficiently threatening or alarming character utilized, or painful cutaneous stimuli employed, and in practically all instances a response or reaction will be evoked, proving the inaccuracy of the general statement already referred to. If the hand be suddenly passed across in front of the patient's eyes with a threatening gesture he blinks and may withdraw the head; at the unexpected bursting of an air-filled paper bag he also blinks; when the auditory meatus, or the nostril, is tickled with a paper spill the head is inclined away and so forth. Only in exceptional instances do such reactions fail. Thus I have seen tears running from the eyes of a Parkinsonian when his anterior nares were tickled, yet no outward reaction was otherwise noticeable; another patient blinked his eyes when a feather entered his external auditory meatus but the head did not move away. I have convinced myself, further, of the general inaccuracy of the view according to which patients with paralysis agitans furnish no visible evidence of a reaction to painful cutaneous stimuli; on the contrary, withdrawal from the nocuous excitation in my experience rarely fails. If the fauces are irritated with a

paper spill moderate "gagging," with movement of eyes and mouth, results more or less regularly.

In seeking an explanation for the occasional absence, and less infrequent reduction, of movements of reaction and defence we do not require, as indicated above, to go past the factors already enumerated, in particular, that of awareness, the action of which has been sufficiently demonstrated. With certain patients of the hospital class the rôle played during examination is so passive as of itself to tend to reduce external reactions to a minimum. Many patients by no means as generally immobile as the Parkinsonian will submit to repeated pin-pricking in sensory tests with stoical calm, and the same is true of ordinary sole-stimulation in investigating the plantar reflex. As a persistent background in rigid cases is the muscular state, which of itself, as we have several times had occasion to remark, contributes to reduction of facile reactions, to an extent, indeed, that cannot be ignored.

Movements of Expression

Hitherto our concern has been exclusively with the voluntary motor system and its activities when there is reason to believe the striatal system is largely out of action, but I must now refer to another class of movement which, commonly called involuntary, is nevertheless more conveniently discussed at this stage than later, when we come to examine chorea, tremor, and athetosis.

Scarcely any greater clinical contrast can be imagined than between the mobile and facile emotional expressions of the Sydenham's choreic and the "starched" and fixed facies of the Parkinsonian. In the latter it is not an absence of all expression but rather a fixity of expression, a lack of ease in passing from one expression to another, with which we have to deal.

Though not specifically alluded to by James Parkinson, the Parkinsonian mask is a classic symptom, not requiring elaboration by any writer of to-day. I may state briefly that patients can and do both laugh and cry, that their expressions frequently exhibit an exaggeration amounting almost to a *rire* or *pleurer spasmodique*, and that in unilateral cases the affected side moves more slowly than the other in the expanding or unfolding of its expression, but tends to maintain this after the other has become normal again. A distinction is to be drawn, none the less, between the true *rire et pleurer spasmodiques* of the pseudobulbar and the exaggerated facial emotional expression of the Parkinsonian, in that the "con-

vulsive" element of the former is wanting in the latter instance, in which also there is far less implication of the respiratory part of the mechanism for the display of emotion, while the quality of the laugh is often inferior. As a postencephalitic patient remarked, "I used to have a ringing laugh, but now I make a silly noise." It is not that, as in pseudobulbars, double hemiplegics, cases of disseminated sclerosis, and so on, the faciorespiratory emotional apparatus is released into exaggerated action (see page 286), but that the apparatus itself, an involuntary mechanism, is subject to the same muscular disabilities, so to speak, as have been described in the case of the voluntary motor system. For we are dealing, as is immediately understandable, with skeletal, not with unstriated, muscles; the problem pertains to the involuntary use of a voluntary musculature. Now there is no good reason to suppose the inward or central (psychical) part of the mechanism has undergone a change; the Parkinsonian is no less conscious than the normal person of the stimulus that conduces to laughter or awakens tears; hence such abnormalities as he shows must be set down, *inter alia*, to the existing hypertonic condition of the muscles involved, that rigidity which seriously interferes with facility in or versatility of muscular play. The important point that I wish to make at present is that *the mimic or expressional movements under consideration are "most automatic" or "least voluntary," but they nevertheless reveal disorder of function as definitely as do ordinary voluntary movements.*

Poverty or lack of movement in gesture and expression is, of course, a mere corollary of what we have learned as to the "Bewegungsarmut" in general. The "mimische Starre" or mimic fixity of the Parkinsonian, inclusive both of facies and of limbs, is a consequence of more than one factor, already sufficiently discussed, and is, in my opinion, erroneously described as a mimic "paralysis," for reasons given above. Hence, when Binswanger and others argue from the symptomatological to the physiological, and place in the corpus striatum a "higher co-ordination centre for unconscious mimic and gesture play," they misinterpret the clinical phenomena in the first instance and overlook other significant clinico-anatomical data in the second. For elsewhere (see page 287) I have brought forward evidence to suggest that there are corticofugal (non-pyramidal) paths from frontal lobes via optic thalamus to faciorespiratory centres in pons and medulla, and that these are the routes taken by emotional impulses to activate the faciorespiratory synkinesis in the direction either of laughter or the

reverse; genuine absence of emotional movement occurs from lesions situated *ex hypothesi* anywhere on these tracts, from the cortex to at least as low as the pons. *No theory attributing emotional disorders to disease of the corpus striatum alone can be accepted, since it fails to include a number of germane clinical and pathological facts.*

The Lore of the Antagonists, and their Rôle in Striatal Disease

I have left to the end of my necessarily somewhat long analysis of movement the vexed question of the nature of the part played by the antagonists in normal voluntary motion, and the correlative problem, whether among motor symptoms of striatal affections any, and if so, which, are to be ascribed to disordered function of this component of movement.

In his Croonian lectures (1903) the late Dr. Charles Beevor¹⁰ summarized the two divergent theories then in vogue as to antagonistic function: (1) According to Winslow (1756) and Duchenne (1867) the antagonists take part in all voluntary movements, their function being to moderate (check, control) the contraction of the prime movers or protagonists, for the sake of precision of movement. Demeny (1890) drew a distinction between resisted and non-resisted voluntary movements; in the case of the former, the antagonists relax; in the case of the latter, they are in action during the primary movement. (2) According to Sherrington, when contraction of the principal muscles taking part in a given movement is effected by cortical (electrical) stimulation there is both relaxation and inhibition of tone of the antagonists of this movement. Even with minimal excitation of the motor cortex relaxation of antagonists is still obtained. From his own clinical observations Beevor concluded that in strong movements against resistance the antagonists are always relaxed, and that in unopposed movements (gravity being excluded) relaxation is the rule.

Needless confusion has arisen from failure to appreciate the exact meaning of "relaxation" of an antagonistic group when protagonists contract. This has been taken erroneously to imply constant, complete, and immediate relaxation, whereas the words of Sherrington are that the contraction of the one muscle rises as that of the other falls, and the rise of the one keeps sufficiently close step with the fall of the other. No primary conflict between the

two theories exists, nor any insurmountable difference. Amplification, however, is desirable.

To get a clear idea of antagonistic function we must bear in mind the *essential variability of antagonistic rôles*. Any stereotyped notion of antagonists being somehow opponents is fatal to clear thinking. We may speak of the antagonists *in* a movement, but never *to* a movement. "Opposite" muscles are not ipso facto "opposing" muscles. Their function ranges from (1) active co-operation in a movement, so that it is indistinguishable from that of prime movers, (2) through harmonious decontraction, as in ordinary displacements of a joint, to (3) a relaxation as quick and complete as possible, when the prime movers are acting powerfully against resistance. In illustration of this latter function, let the observer extend his arm laterally to a point well above the horizontal level at the shoulder, the deltoid being in full protagonistic action. In this position let him place his hand on a convenient shelf or ledge and make strong downward movement of the extended arm against this resistance. No displacement is possible, but the deltoid, now the antagonist in the required movement, immediately and with remarkable completeness can be felt under the fingers to soften and relax. There is here no moderating function, since no displacement as a fact occurs. In illustration of the first-mentioned function, I may cite the "straight left" of the boxer, when, accurately speaking, protagonists and antagonists as usually understood are in active co-operation, there being again no joint displacement. Beevor's illustration of the conversion of the leg similarly into a rigid muscular pillar, as when skating on the outside edge, is equally apposite.

As I conceive it, recognition of this variability in antagonistic action will explain what might otherwise appear mutually discordant phenomena. At any moment in the course of a voluntary movement the character of antagonistic co-operation may alter according to requirements; it may change from decontraction to synergic co-operation, or again to relaxation, as the prime movers pass from kinetic to static action, or encounter resistance, and so forth. Differing modalities of function in the antagonists correspond to variations in protagonistic action. No haphazard or indiscriminate correlation, however, exists between the two; on the contrary, all is in strict physiological harmony.

As is known, the relation of agonist contraction to simultaneous antagonist decontraction in ordinary joint displacements was

described by Sherrington as reciprocal innervation, and this law has received unqualified acceptance ever since its enunciation. That any conflict of the "pull-devil-pull-baker" kind should exist as between, say, flexor and extensor groups during displacement at the elbow is both clinically and physiologically unthinkable. He also, however, has demonstrated the occurrence and explained the nature of what he calls double reciprocal innervation, a physiological state in which the centres for both prime movers and antagonists may discharge together, though unequally. In this way Sherrington covers the cases both of increase of protagonistic contraction with diminution of antagonistic contraction, and of simultaneous contraction (or, of reversion into rest) of the two groups.

Tilney and Pike,¹¹ however, have recently come to other conclusions. These investigators, experimenting on cats and monkeys, state that "under normal conditions we have not observed the contraction-relaxation phenomenon which is generally known as Sherrington's 'reciprocal innervation.' This phenomenon, although also observed by Beaunis in rare instances, was deemed by him both exceptional and inexplicable." Their view is that in all voluntary movements synchronous "co-contraction" takes place in the antagonists; they speak of "synergic units" composed of both flexors and extensors, and so on, and in each "synergic unit" there is a "dominant" element (the prime movers) and a "check" element (the antagonists). During action each exhibits "a proportional mutual increment of tension," which in the case of the former is "uniformly greater" than in the case of the latter. There are also differences in the "times" when this increment of tension comes into action in the two respectively. I cannot in this place attempt any examination of their researches as these may bear on the problem before us, but I may allow myself one comment. These workers do not appear, on the clinical side, to have covered all the possibilities of antagonistic action, since no investigation, seemingly, either of voluntary movement against strong resistance, or of antagonistic co-operation, has been made. The experimental side of their research undoubtedly seems to conflict on the face of it with Sherringtonian conceptions based also on experimental methods, but I do not think there is so complete a disparity as might at first appear between some (at least) of their findings and the usual clinical interpretation of the rôles played by muscular components in individual voluntary movements.

Relying, then, on well-tried clinical procedures and their application in the study of striatal disease, my conclusion is that *in these affections the general laws of antagonistic action, as above illustrated, hold good*. From the records of Parkinsonian cases reproduced in Figs. 16, 17, and 19, contraction and decontraction can be demonstrated just as in normal subjects. No "defective inhibition of antagonists," that is, no marked retardation, or failure, in making a particular movement on request through inability to relax the existing tension in an antagonistic muscle, has been noted, still less any instance of the antagonists coming into action *before* the prime movers, as we shall see occurs in other pathological conditions to receive attention. The Parkinsonian can employ his antagonists in co-operation, and, conversely, they can be shown to slacken when the occasion demands it, according to normal physiological precedent.

I find, of course, as has been already outlined, an impediment to quick alternating movements of a limb-segment in the existing hypertonia of *both* of two opposite groups (see below, page 195). It must be obvious even to superficial observation that the fingers cannot be separated with facility or quickness by one set of inter-ossei if the reflex tonic contraction (hypertonia) of the opposite set cannot be inhibited with facility either. For while volition *adds* a contractile element to a muscle already in involuntary contraction, in the sense that the two blend, its action in the antagonistic component, also in involuntary contraction, should be to allow the latter to elongate and yet to maintain in elongation a moderating influence requisite for the particular end to be served. Admitting antagonistic activity in joint displacement, and not a mere passive or mechanical stretching of a relaxed muscle, the truth remains that this activity of innervation, this "increment of tension," is occurring in a muscle that is in fact *lengthening* itself, and the question is to explain the simultaneous activity of contracting and elongating elements. Only by inhibition, as I conceive it, of a *shortening* contractile process (such as takes place in the prime movers) can a muscle actually elongate while maintaining contractile tension; that is to say, the process in the antagonists cannot be physiologically *identical* with that in the protagonists. When a muscle is hypertonic its fibres are in a condition of greater (reflex) contraction than in a normal muscle. If it happens to be *pro tem.* an antagonist, then decontraction starts as it were behind scratch, and is adversely handicapped from the outset. Avoiding

the language of metaphor, I say that more decontraction has to be effected in the antagonist (by a process of what I can only regard as inhibition) before the muscle begins to elongate, than is requisite in a normal antagonist. Hence the hindrance already alluded to, an impediment traceable through all Parkinsonian action in greater or lesser degree, and one to which many of the clinical phenomena here illustrated are in part at least attributable. Hence, in part, also, the "effort" of which the Parkinsonian patient is for ever complaining. The amount of innervation required for everyday movements, familiar to him by years of experience, turns out to be insufficient for the desired purpose; and he utters the simple confession, "it is easier not to make the movements."

An important and significant corollary arises from the conclusion to which this clinical study of striatal cases has led. Having found no abrogation of agonist-antagonist activities, no material departure from normal physiological laws except in so far as existing hyper-tonia causes a drag on rapidity and a reduction of facility, I am led to the decision that *the normal combining of protagonist, synergic, and antagonist muscular units in a single voluntary movement cannot be regarded as a function of the corpus striatum*. I need not at this point enter on a discussion as to the physiological level at which this combination takes place, since it will be dealt with subsequently. Suffice it to say that since the lesions of the cases under review are in all probability destructive or degenerative processes in corpus striatum and its efferent systems, and since, as I have just pointed out, muscular components are in normal (if sometimes hampered) action as required, the mechanisms underlying their action are definitely non-striatal. Statements such as that of Lewy,¹⁹ therefore, that "the slackening of the antagonists during contraction of the agonists is dependent on the corpus striatum," are without either clinical or physiological foundation.

(For references, see list at end of Chapter XI.)

CHAPTER IX

DISORDERS OF MOTILITY AND OF MUSCLE TONE, WITH SPECIAL REFERENCE TO THE CORPUS STRIATUM

III. *DISORDERS OF MUSCLE TONE IN STRIATAL DISEASE.*

Introduction. Distribution of Muscle Hypertonia. Its Characters. Plasticity. "Lengthening and Shortening" Reaction. The "Cogwheel" Phenomenon. Striatal Attitude. Criticism of Hunter's Views on Parkinsonian Tone. "Fixation-contraction."

Introduction

By muscle tone is commonly understood that condition of contraction of a resting muscle which exists without voluntary innervation and which holds the limb in a given position. It also exists when the limb is passively supported, for the muscles do not sag or hang flaccid. When, further, the limb is passively moved a mild resistance to displacement is encountered; when it is held and shaken the segments concerned exhibit no flail-like swinging as do those of a limb considered to be hypotonic or atonic. Thus muscle tone gives shape to muscles, and is in evidence both in resting (static) and moving (kinetic) states of the musculature. According to Sherrington, tone is in reality postural contraction; that is to say, it is a moderate contraction of muscle-fibres effected reflexly by sensory excitations arising in peripheral end-organs, substantially those of the muscles themselves. In virtue of this reflex contraction, a muscle will adapt itself to the maintenance of any posture, whether the fibres are as a fact in a shorter or longer anatomical state; the property of adjustment of length of fibre, and of exercise of contracting power whatever the length, he described as "plasticity," whence has arisen the use of the term "plastic tone," to be referred to immediately. All clinicians agree that lesions interrupting the proprioceptive reflex are underlying

muscle tone confirm the conception by producing, as a fact, loss of tone.

The point next arises whether this muscular function of reflex contraction is subserved by all or by a special group of striated muscle-fibres. I can now only refer in the briefest fashion to a hypothesis that has gradually developed more particularly during the last decade, to the effect that all fibres partake in postural tone but that (as the form of the hypothesis due especially to Hunter²⁰ has it) there is a subdivision of labour, so to say; some fibres, said to be histologically recognizable, are innervated by the motor or efferent limb of the proprioceptive arc alluded to above—i.e., by the somatic nervous system—and contract reflexly during change of posture; they are considered to be responsible for what has been called “contractile tone,” because they contract in response to appropriate stimuli. Other fibres, stated to be histologically distinguishable, are innervated by the sympathetic nervous system but they are not contractile; their length is supposed to be *passively* altered during the activity of neighbouring contractile fibres (the first group); and their postulated function is to remain at a given fixed length once movement of the others has ceased, and thus to aid or support the maintenance of posture determined otherwise. They are said to be responsible for “plastic tone”—i.e., the element in postural tone concerned with the adaptation of a muscle to the new posture assumed after movement. “Plastic tone” and “contractile tone” together make up “postural tone.”

While Hunter asserts the sympathetically innervated fibres for “plastic tone” are *non-contractile*, he makes no reference, as far as I have seen, to the old Vulpian’s phenomenon (1862), recently reinvestigated by Langworthy.²¹ The latter finds that stimulation of the lingual nerve (containing bulbar autonomic fibres) leads to slow wavy contractions of tongue muscles after complete degeneration of hypoglossal nerve-endings by previous section, and is inclined, therefore, to believe in the possibility of *contraction* via the sympathetic nervous system.

Be this as it may, the identity of tone and of contraction cannot be seriously questioned. For physiological proof of this statement the clinician has to turn to recent work with the string galvanometer. Very briefly, it has been shown by exact methods that an action-current exists in normal muscle at rest, that there is no notable difference in the action-current as between active (“voluntary”) movement and passive movement, that the action-current

of normal muscle differs from that of rigid and spastic muscles only quantitatively, not qualitatively. Further, the curves of the current during passive movement of a hypertonic muscle are the same as those of strong voluntary innervation; no qualitative distinction is to be found between action-currents (during both active and passive movement) of Parkinsonians and of hemiplegics; or between action-currents of the same conditions when muscles are at rest. In respect of this latter point, the curves both in hemiplegia and in paralysis agitans are more marked than in resting normal muscle, and in paralysis agitans rather more than in the pyramidal affection (Hansen,²² Hoffmann,²³ von Weizsäcker,²⁴ Wachholder,²⁵ Mann and Schleier,²⁶ and others). The general conclusions are, that tone and active innervation are analogous phenomena, that only quantitative differences are to be detected between action-currents of voluntary innervation and of hypertonic states of pyramidal and extrapyramidal origin, and that the muscle tone exhibited on passive movement in these morbid conditions (resistance to stretching) is analogous to the tetanic innervation of voluntary contraction. A number of years ago Lewandowsky²⁷ maintained that "tonus is nothing else than the observation of a particular attitude for which the same laws hold good as for the carrying out of a movement."

What actually happens when volition innervates a resting tonic muscle is probably that it strengthens the existing (involuntary or reflexly produced) contraction, as is suggested by a significant experiment of Graham Brown, to which attention is drawn elsewhere (see page 129). According to Holmes,²⁸ tone "fuses with muscular contractions, otherwise aroused [e.g., by 'volition'], reinforcing them, making them more tonic." The reader is referred again to the tracing obtained from the erector spinæ of a Parkinsonian during the combined arm movement described above (Fig. 15). From this record it is clearly established that the existing involuntary or postural contraction of that muscle (its tone—in this instance, hypertonia) is *reinforced* by the volitional element of active contraction, so that the curve rises still further, if somewhat slowly. I attach importance to this demonstration, since Hunter asserts (without, seemingly, offering evidence in support) that "in voluntary movement postural tone, made up of a contractile and a plastic component, is first *inhibited*" (italics mine); according to my observations this is incorrect.

The difficulties of gauging tone by ordinary clinical methods are

considerable; nor are they diminished by inability to separate voluntary from involuntary contraction objectively, or by the fact that the patient often unintentionally co-operates during passive movement. While major degrees of hypo- or hyper-tonia do not, as a rule, offer any difficulty, minor degrees escape us clinically, and the passing from a reflex contraction to an incipient voluntary contraction (without displacement) is for practical purposes not determinable. Conversely, we lack methods of gauging with exactitude at what point a postural tone-reaction fixes or stabilizes a movement begun by volition.

A further introductory word is necessary in respect of the recent tendency to differentiate somewhat schematically movement from posture, and kinetic from static muscular contractions. Speaking generally, volitional action is taken to imply *movement*, and reflex action, *posture*, yet the inaccuracy of the generalization is plain; posture is often maintained voluntarily, and much reflex action is kinetic. Movement is in reality a series of changes of posture, and Sherrington has stated that the distinction between reflexes of posture and reflexes of movement is not clear-cut. That two *distinct* mechanisms should be responsible for posture and movement respectively is *prima facie* improbable, in view of this statement of Sherrington's for reflex action, and of the known phenomena of voluntary action. Cogent arguments against this differentiation have been urged by Bard,²⁹ with whose contentions I find myself in substantial agreement.

Muscle Tone in Striatal Disease

Study of an average case of Parkinsonism or of progressive lenticular degeneration, even by the comparatively crude clinical methods to which we have resort, at once reveals the presence of increase of muscle tone. The muscles stand out and have shape; where their tendons are visible these are salient; to the palpating finger they are firm and tense; they offer definite resistance to passive extension, and more force is required to effect passive displacement. Further, such patients exhibit at rest an attitude indicative of access of tone in certain groups.

DISTRIBUTION OF MUSCLE HYPERTONIA

The bellying of the skeletal musculature is more or less universal, but is more recognizable, on the whole, in those groups whose contraction underlies the maintenance of specific Parkinsonian

attitudes (sternomastoids, erectores spinæ, deltoid, biceps brachii, supinator longus, wrist extensors, long finger flexors, interossei, thenar and hypothenar groups, knee flexors, tibialis anticus, etc.). Rigidity—i.e., resistance to passive movement—is so widespread that it may be described as general, though it is more pronounced at the larger joints, being presumably there a question simply of muscle mass. Even when a limb or limb segment is passively put in such a position as to relax muscles as far as possible hypertonia is evident. In the recumbent posture the sternomastoids do not relax. When any joint of any limb is passively displaced resistance is encountered whatever the direction of the displacement. We have already had evidence pointing to the implication of small muscles—e.g., those of the larynx—in this generalized rigidity, and it is doubtful whether external ocular muscles completely escape. In the result, the patient can often be passively moved *en bloc*; if the head be rocked about the trunk follows its every move, and so on. A degree of rigidity equal to that of the severest pyramidal cases is probably attained but seldom (unless, indeed, the stage of myogenic contracture has been reached); it is not so easy to pull a Parkinsonian about by one leg, the other following, as it is in the case of a pronounced spastic paraplegia.

CHARACTERS OF THE MUSCULAR HYPERTONIA

When the hypertonic muscle is passively stretched resistance is felt from beginning to end of the movement; in mild cases its origin and insertion points should first be approximated and the passive stretching should be comparatively fast; if slow, little or no resistance may be demonstrable. The explanation doubtless is to be sought in stimulation of proprioceptive end-organs by abruptness of initial passive movement, with consequent reflex increase of muscular contraction and therefore of resistance. Strümpell is of opinion that striatal rigidity remains more or less the same whether passive movement be slow or abrupt, gentle or powerful, but my observations do not confirm this view, nor do I consider it can be applied as a clinical method of distinguishing pyramidal from extrapyramidal rigidity. Figs. 21 and 22 are two records from the same muscle (biceps brachii) of an advanced postencephalitic Parkinsonian; in the former of these the muscle (already contracted) was passively and slowly extended in the course of three seconds; in the latter, the same movement was made abruptly and strongly, occupying not more than a quarter

or a third of a second, and the characteristic "cogwheel" character of the resistance stands out with greater clearness in it than in the other.



FIG. 21.—Parkinsonism. Slow passive stretching of biceps brachii.

The statement has already been made that rigidity is practically universal in striatal cases; in the various divisions of the musculature it varies in degree only. It is found in muscle groups which in pyramidal affections exhibit little or none.

Like one or two other phenomena of postencephalitic Parkinsonism, rigidity may show a curious reversibility, fluctuating from time to time in an unexpected degree—a character not met with to the same extent in pyramidal cases. In deep sleep careful observations by my house physicians have proved it to be markedly reducible, frequently if not constantly. Cutaneous sensory stimuli exercise no apparent effect on its character; it is not temporarily broken down thereby, in contradistinction to what obtains in pyramidal disease. It seems to be aggravated by cold and reduced by warmth.

In spite of the plausibility of the view that muscle rigidity is mainly responsible for impairment of voluntary movement, I am not able to satisfy myself there is constantly a specific connexion between the two. I have not found that movement is always braked by existing rigidity and brought to a standstill more or less completely and soon, as Foerster states. Peripheral rigidity is only one of the factors producing limitation of range and power,

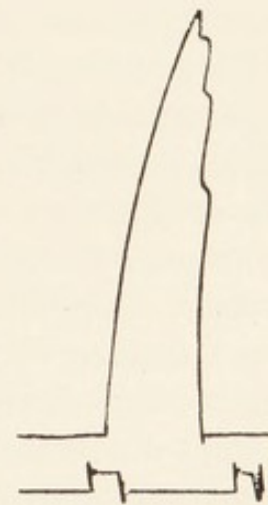


FIG. 22.—Same case as Fig. 21. Quick passive stretching of biceps brachii; "cog-wheel" phenomenon well seen.

and constituting paresis, in respect of voluntary contraction (see page 160). I have demonstrated the physiological normality of the mechanism of volitional innervation in regard to its muscular components, concomitant rigidity notwithstanding. We have had evidence to show that voluntary muscular contraction blends with existing tone and is rendered more tonic. Were rigidity really to function as a constant and immediate brake on volition a difference might be expected between the curve of a rigid muscle under passive and active extension respectively, since in the latter the extension is part of a volitional action. By physiological (action-current) methods no such difference is discoverable. With cruder clinical technique the arm of a patient with severe paralysis agitans was fixed so as to prevent any subsidiary movement; the biceps brachii was in the first instance passively stretched, the time taken being roughly two seconds; another tracing gave active ("voluntary") extension of the same muscle, in the same time, and under the same conditions, and no distinction could be found. However, single instances must not be unduly stressed, and from this illustration, further, the inference is not to be drawn that hypertonia does not impede passive movement, for the reverse is the case. In analysis of the actual hindrance encountered by volitional impulses in the motor periphery due weight must be placed on the state of the antagonists, as we shall shortly see. This is of greater significance than hypertonia of the prime movers, though one might conceive so exaggerated a condition of agonist hypertonia that the incoming current of volitional innervation produces little or no evident result. No such case, as a fact, has as yet come under my notice.

A further word is necessary in this connexion as to the type of movement influenced by abnormal states of the musculature. From observations made in the course of testing various kinds of movement, as described previously, the element of rigidity has been seen to contribute to the general imperfection in all; that is to say, it hampers movements of a "most automatic" kind no less than those that are "least automatic," as might, indeed, be expected. If there is a persisting background of agonist-antagonist rigidity, volitional innervation of a given muscular unit cannot differ from automatic innervation, since both use the final common path in a Sherringtonian sense. I have not found, therefore, in working on this class of case, any support for the contention of Kousnetzov,³⁰ who says that in extrapyramidal hypertonia "it is above all the voluntary movements that are affected."

Plasticity : "Lengthening and Shortening" Reaction

Proof of the existence of the property of plasticity in muscle tone we owe to the investigations of Sherrington, who in his work on experimental decerebrate rigidity discovered that the limb of the animal, when passively flexed, say at the knee, would retain that posture, the stretched quadriceps adapting itself to the new position of the limb and holding it so ("lengthening" reaction); conversely, if the latter was again extended, the quadriceps naturally shortened, yet again contributed its quota to maintenance of the fresh attitude ("shortening" reaction). We note that in Sherrington's view this quality belongs to tone as derived entirely from somatic proprioceptive arcs, and that it is demonstrated only in pathological (decerebrate) states, in which one need scarcely say various efferent paths *both* from the cortex *and* the basal ganglia are completely out of action.

In the course of study much attention has been directed to the question of plasticity, and the general conclusion is, that *this quality of muscle tone is not specially in evidence in striatal cases*. Parkinsonian limbs, passively compelled to assume a certain posture, reveal no particular tendency to maintenance of the new position. On the contrary, when left to themselves they tend to return forthwith to the Parkinsonian attitude from which they have been disturbed, that is, if the case is of average severity. A patient told me spontaneously of his frequent observation when in bed that on his extending his arms outside the bedclothes they always came back insensibly to a position near the upper part of his chest. This return to previous attitude is easily observable in the case of the hand and fingers; passive extension of the fingers is not succeeded by maintenance of the extended posture, even for as short a time as a second or two, whereas in decerebrate preparations the fresh posture is held almost indefinitely.

The feature of maintenance of different postures passively imposed on a limb is characteristic of the clinical state known as catatonia, and is not in itself dependent on the presence of extremes of rigidity. Catatonia is seen in a considerable number of morbid clinical conditions, diverse in nature but alike in that cortical function is disordered. It is often noticed in the acute stages of epidemic encephalitis, in various psychoses, in certain cases of hysteria, and so on. In respect of *outward* character, the "lengthening and shortening" reaction of a decerebrate limb does not differ

from that seen in any of the above-mentioned morbid states, in which, precisely, the limb muscles, in virtue of plasticity, hold it in any posture passively impressed on it. The argument is not vitiated by any claim that in these cortical and psychical conditions the action of volition is not excluded, for investigation will prove the claim to be unfounded. Now in my experience *no clinical state resembling catatonia, even if merely in external characters, occurs in ordinary cases belonging to the striatal group.* Applying usual clinical tests, the observer finds little evidence of the occurrence of any reaction to be appropriately described as a "lengthening and shortening" reaction, such as Walshe,³¹ for instance, has had occasion to note in a case of decerebrate rigidity in man, and Royle,³² in spastic paraplegia of cortical origin (gunshot wound). I cannot therefore give support to the view of Hunter,²⁰ as expressed in his latest articles, that "a tendency for fixation of the muscle at any length passively imposed upon it" is a special characteristic of Parkinsonian rigidity. On the contrary, the cases in which this has come under my notice have been mainly extrastriatal. In view of its interest, allusion may be made to a recent instance.

The patient was an elderly man who was admitted to hospital in a semi-conscious state, with a history of having had a stroke twenty-four hours previously. On examination the right arm was seen to be in a state of "early rigidity" and in moderate flexion; the right leg was similarly rigid and extended. Aphasia was prominently in the foreground of the clinical picture. The man frequently passed his left arm over his head or moved it spontaneously about the bed, whereas the right limbs exhibited no such movements. The deep reflexes were more active on the right than on the left; the plantars were both in reduced flexion, and a crossed plantar in flexion was constantly obtained from the right sole, but never vice versa.

It was remarked at once on testing that the right limbs (arm and leg) exhibited the "lengthening and shortening" reaction in a pronounced form; as a single illustration, when the patient was put on his left side, the right lower extremity was passively abducted at the hip and flexed to a right angle at the knee, and this new (and awkward) posture was maintained for three minutes by the watch. The leg was then extended at the knee, and the quadriceps group at once maintained the extension again. The left limbs showed a similar reaction in a less noticeable degree; it was usually and more or less immediately interrupted by the spontaneous movements already alluded to.

On the second day the patient had two typical Jacksonian fits on the right side, beginning with twitching of the right arm, and both of these spread in moderate degree to the "second side." A diagnosis of cortical (submeningeal) hæmorrhage over the left frontoparietal region was made and this was corroborated by subsequent examination.

A case of this kind confirms the general statement made above, to the effect that the type of clinical case most likely to furnish proof of the existence of plasticity as a quality or property of muscle tone is one in which *pyramidal* function is disordered from the cortical side.

Passing reference may at this point be allowed to the experimental evidence on which Hunter ²⁰ partly relies for support of his views. In the domestic fowl and seagull he removed the corpus striatum on one side and obtained a diffuse muscular rigidity, which he says "is remarkably similar to Parkinsonian rigidity in man. Its diffuseness, resistance to stretching and the tendency for fixation of the muscle at any length passively imposed on it, are common to the two conditions." Now it may be fairly argued that the bird is not a suitable animal for experiments from which deductions are to be drawn as to striatal function *in man*, for the plain reason that it has practically no cerebral hemisphere beyond the corpus striatum; its pallium is electrically inexcitable; it has no pyramidal tracts in the ordinary sense; in its tractus striomesencephalicus (spared by the operator) mingle fibres both of striate origin and from the minute extrastriate pallium. The differences between it and the mammalian brain are great enough to render comparisons as to function more than usually precarious.

Since in the sense employed by Sherrington the reaction of plastic tone is a property of ordinary somatic proprioceptive tone, and since hypertonia characterizes paralysis agitans and the other diseases at present under discussion, we might expect to be able to demonstrate at least minor degrees of the lengthening-and-shortening reaction in these affections. Well-recognized distinctions, however, between pyramidal and extrapyramidal cases must be borne in mind; prominent here is the fact that nothing like the same diminution of voluntary innervation obtains in the latter as in the former. Speaking generally, the subject of paralysis agitans is capable of innervating his musculature to an extent impossible for the patient whose corticospinal tract or tracts are anatomically or physiologically interrupted, and for this reason, as I consider, the lesions in his case cannot produce the pathological conditions essential for visible demonstration of the phenomena of plasticity in any marked degree. In other words, the more serious the interruption of the pyramidal system, the more easily is this particular tone-reaction excited; and it is seen at its maximum when that system is completely in abeyance, as in decerebration.

The "Cogwheel" Phenomenon

We may now consider whether the "cogwheel" symptom of Camillo Negro (1901) is connected with the reaction in question, and whether it may be regarded as a minor degree of the lengthening-and-shortening sign. When a hypertonic muscle is passively stretched it resists, and this resistance often takes the form of an irregular jerkiness, as though the muscle were being pulled out over a ratchet arrangement; it is momentarily checked and extended in a little alternating series of two, three, or more phases, hence the expression imagined by Negro. If the passive stretching is quick the phenomenon is usually more readily appreciated by the examiner. (Cf. Fig. 22.) It is not found in the opposite muscle that is being simultaneously shortened; on active movement it may be remarked both in contracting and relaxing muscles, but to a less extent than in the case of passive movement.

Various and very different explanations have been forthcoming. Santos,³³ for instance, an exponent of the dual theory of muscle tone, ascribes it to a clonus of the parasympathetically innervated part of muscle, but if Hunter's opinion that this part is compact of non-contractile fibres be adopted it is at once obvious these cannot of themselves be the seat of any such movement, for *ex hypothesi* they are only passively moved with the contracting fibres. Bing,³⁴ who remarks that this view does not account for the occurrence of a degree of the cogwheel sign in active movement, seeks to explain it as a disturbance of reciprocal innervation, a release-phenomenon "produced by failure of a subcortical mechanism which ensures continuity of decontraction." It is simpler, I consider, to regard it as one of the expressions of hyperactivity on the part of somatic proprioceptive arcs. When a muscle already in contraction (hypertonia) is abruptly stretched its end-organs are stimulated and reflex contraction promptly reinforced, thus offering a momentary increase of resistance, felt as a sort of "catch" during passive extension; the moment this "catch" takes place there is an immediate tendency to fixation at that point, for the muscle, being in a condition of postural overtone, will exhibit this tendency whenever it is momentarily "pulled up" (movement being the process of changing from one posture to another). In its turn the "catch" is overcome by the examiner's effort and the stretching causes a second excitation of the arc, hence again the process is gone through. If a muscle, however hypertonic, is slowly pulled out the phenom-

enon is so faint as to be negligible. In a sense, therefore, the cog-wheel phenomenon may be regarded as a minor form of the lengthening-and-shortening reaction. It is not, however, confined to Parkinsonian and similar cases, though naturally more readily there demonstrable because the muscle can in fact be stretched; in severe hemiplegic cases the rigidity may not be overcome by stretching, but when it can be the observer is sometimes conscious of a mild cogwheel movement. Its appearance in the course of active or voluntary movement of a rigid motor unit (in protagonists and antagonists) is less noticeable than during passive movement; I have not been able to satisfy myself that it is much in evidence in voluntary shortening, but it may be occasionally felt in voluntary lengthening in Parkinsonian cases, where the explanation cannot be essentially dissimilar. If denervation or decontraction is not prompt enough the transient check brings about the same result because of the hypertonia.

Striatal Attitude

So familiar and definite are the attitudes adopted in average cases of Parkinsonism (of whatever origin) and of progressive lenticular degeneration that no detailed account is required. Parkinsonian patients, young or old, all look like each other, mainly because of involuntary assumption of more or less identical postures, which appear to be as specific as those of hemiplegia. The attitude is one of generalized flexion, as a rule; the lower extremities are as it were folded in moderate flexion at ankle, knee, and hip; the trunk is flexed in a concave anterior curve; the head is commonly flexed; the upper limbs are semiflexed at the elbows. At the wrists, however, extension ordinarily occurs, with flexion at the metacarpophalangeal and usually also at the interphalangeal joints. Occasionally, as is not well enough known, a type of Parkinsonism with extension of head and trunk is met with.

Posture or attitude cannot be considered apart from tone-affecting mechanisms. Tone is posture; that is to say, in toneless limbs no particular posture can develop, except as caused by extrinsic agencies—e.g., gravity. Limbs whose musculature is in tone (reflex contraction) assume postures capable in the normal person of being modified instantaneously at the bidding of volition, whereas in disease (pyramidal and extrapyramidal) tone-postures are a sequel to release from the control of superior, tone-controlling centres. The postural scheme of the hemiplegic is no haphazard thing; a

predilection type or pattern exists, possible explanations of which are mentioned in another Chapter (see page 131). In decerebration, analogously, a selective incidence of tone on the antigravity muscles takes place. These specific postures can only be assumed if at the same time the inflow of tone to antagonistic groups is relatively lessened by reflex inhibition. In the case of hemiplegia, tone influx is divided; in the arm, the flexor groups are hypertonic, in the leg the extensors. It is interesting and no less important to note that in each instance the groups with lessened tone are the weak muscles (e.g., the flexors, in the leg). From what has already been said of the blending of tone with muscular contractions otherwise evoked this can be readily appreciated.

Now in the case of Parkinsonian posture the circumstances differ somewhat; there is not the same "schizotonia" (to borrow a useful term coined by Dusser de Barenne) as in pyramidal and decerebrate conditions. The feature of paralysis agitans and kindred morbid states, from this standpoint, is the apparently indifferent or diffuse distribution of the hypertonia, as has been made the subject of allusion already. Yet if the increase of tone were *strictly* equal in each and every skeletal group of prime movers and antagonists, I do not see why resumption of postures as definite and specific as the Parkinsonian should take place after they are passively or actively disturbed. We are forced to the conclusion that, general hypertonia notwithstanding, it preponderates in flexor as opposed to extensor muscles; more precisely, in those groups (already specified) whose contraction underlies the maintenance of Parkinsonian attitudes.

On examination these muscles are seen to be firm, to have shape, and to be outlined in contraction with a distinctness as manifest as in hemiplegic groups associated with posture, and I cannot therefore agree with Hunter that in Parkinsonian rigidity there is no selective incidence, or with his corollary that only contractile tone (in his sense) is thus selective. Nor can I confirm his view that "the posture assumed in Parkinsonian rigidity is due to plastic tone *only* (italics mine), exhibited in agonists and antagonists." On his own hypothesis plastic tone is due to *non-contractile* fibres moved passively by contracting fibres and subsequently held thus in posture, hence the latter must first be in activity, and plastic tone of itself can do no more than support a tone-scheme previously and otherwise brought about. To exclude contractile tone, therefore, in so plain an attitude as the Parkinsonian seems impossible as well

as unnecessary; and if plastic tone is exhibited *indifferently* in agonists and antagonists, the adoption of a specific posture is unexplained.

Of the relative specificity of Parkinsonian attitude there can be, in my opinion, no question, but I can offer at present no satisfactory explanation of its type. No merely mechanical preponderance, as it were, of flexor over extensor contraction suffices. Possibly weight should be laid on the contrast between the relative incidence of tone in decerebrate cases on extensor or antigravity musculature ("reflex standing"), and in paralysis agitans on flexor musculature. But the "meaning" of the latter posture is obscure. In any case, the tone-picture of Parkinsonism is distinct from that of double hemiplegia; yet in decerebrate cases that of the latter is more closely approached than the tone-picture of the former, although the corpus striatum is just as completely out of action (see page 138).

"Fixation-contraction"

Much attention has been devoted, more especially by continental workers, to the question of the occurrence of what is called "fixation-contraction" in cases of striatal disease. The term is not familiar to English neurologists, but the condition has long been known under the name of "Westphal's paradoxical contraction," and consists typically of an involuntary (reflex) contraction of a muscle, of a slow and tonic character, when its ends are passively approximated as closely as is practicable. In suitable instances it can be demonstrated in the tibialis anticus by firm passive dorsiflexion of the foot, and this "tibialis phenomenon" was obtained years ago by Oppenheim in cases of paralysis agitans. In 1906 Foerster stated that "fixation-contraction" can be seen in *all* the muscles in that disease, as well as in cases of arteriosclerotic muscular rigidity. It has been called "fixation-rigidity" by Strümpell. Jakob⁴ attributes considerable importance to the phenomenon, which has also been studied by Delmas-Marsalet.³⁵

In routine examination special attention has been given to fixation-contraction, following the technique of Foerster, who says the passive movement (of shortening) has to be made with a certain jerk and the limb segment pressed firmly to the end-position, the head of the bone concerned being pushed well into its socket, and the new position held passively for an appreciable moment. When this technique is prosecuted the muscle passively shortened exhibits

a tonic contraction analogous to the shortening reaction already investigated as a property of somatic proprioceptive tone.

As a result of tests directed to this matter I have come to the conclusion that *while fixation-contraction can be demonstrated in a fair number of cases it is neither universal in the musculature nor so pronounced as Foerster would have it.* In various typical cases of Parkinsonism I have failed to satisfy myself of its presence except in minor degree, and not seldom, indeed, is it negligible. A further conclusion is, that *it is no more than a manifestation of increased somatic proprioceptive tone and as such is not deserving of a special terminology.*

To investigate its occurrence a number of cases of well-marked unilateral Parkinsonism were selected and the condition on one side compared minutely with that of the other. The tibialis phenomenon was made the subject of special examination; another convenient test is to approximate the origin and insertion of the deltoid by passive lifting and supporting of the arm in the vertical position. Many other muscles, however, were also utilized. To convince oneself of the presence or absence of fixation-contraction the muscle in question was examined with the eye, was palpated with the finger (an assistant performing the passive movement), and, if it so happened that the action of gravity came into play when the limb segment was deprived of this passive support, the time taken to fall was estimated on the two sides respectively. Speaking generally, no pronounced or persistent tonic contraction in the approximated state was observed in any particular muscle—at the most, as already stated, only moderate or minor degrees of the condition could be found.

In no case have I or my assistants been convinced of the development by this method of a muscular state that can fairly be represented as belonging to the category of catatonia (or catalepsy, as both Jakob and Foerster term it, though use of the word in this connexion should be avoided). If the foot takes longer to drop through gravity on the affected than on the normal side, if as the arm falls from the vertical an evident “catch” is demonstrable just as the movement begins, this is no more than what is to be expected in a hypertonic muscle or muscle group, and cannot be thought to constitute per se a pathognomonic sign of extrapyramidal rigidity. There is truth in the argument of Mayer,³⁶ that in some instances at least fixation-contraction is nothing else than a stretching contraction, such as has been already illustrated. Even

in well-marked examples of Parkinsonian rigidity, when passive shortening of a given muscle (as when the flexed lower extremity is extended to an angle, say, of 135° at the knee, the quadriceps being thereby shortened) is made, but the extreme position not attained, it is not my experience that the limb is thus held immobile by the development of a fixation-contraction, or, even so, I cannot assent to the interpretation put on this by the observers mentioned. At the most, it is identical with a phase of the lengthening-and-shortening reaction, a property of proprioceptive tone, and merits neither separate description nor explanation.

When, therefore, Jakob ⁴ maintains that the phenomenon of fixation-contraction is responsible for "cataleptic" (catatonic) symptoms in Parkinsonism, for the development of contractures, for adiadochokinesis and various "pulsions," impeding of voluntary movement, slowing of movement at its commencement, and interruption of movements in series, all this in my view is tantamount merely to the fact that hypertonia (exaggerated reflex contraction) of the musculature interferes with the prosecution of normal muscular action.

(For references, see list at end of Chapter XI.)

CHAPTER X

DISORDERS OF MOTILITY AND OF MUSCLE TONE, WITH SPECIAL REFERENCE TO THE CORPUS STRIATUM

IV. INVOLUNTARY MOVEMENTS AND THEIR PATHOGENESIS. CHOREA AND ATHETOSIS.

Preliminary Considerations. Chorea. Clinical Characters. The "Choreic Hand." The Pronator Sign. Paralytic Chorea. Summary. Athetosis. Clinical Characters. Relation of Athetosis to Tonic Innervation. Pathogenesis of Choreo-athetosis. Conclusions as regards Chorea and Athetosis.

In order to keep our study of involuntary movements within appropriate limits I shall deal mainly with chorea, athetosis, and tremor. At the outset the caution must be repeated, that discussion of these symptoms in the present setting is not to be taken inferentially as implying for them a striatal origin. Further, the reader may with advantage recall the general comments made at the outset on the subject of voluntary movements and on the lack of precision in the connotation of the term.

Preliminary Considerations

Before proceeding to clinical analysis, we must clear the ground by one or two preliminary considerations of basic significance, neglect of which still continues to be a source of confusion.

(1) It seems almost a platitude to point out that the phenomena to be dealt with are "positive" symptoms; they are over-mobile conditions; being movements, they cannot possibly be *caused* by impairment or destruction of movement-producing mechanisms—only paralysis, or absence of movement can thus result. Involuntary movements must be set down to *activity*, not to paralysis, of some or other motor mechanism, which must be in a *physiological*, not a pathological state. In every case of involuntary movement

the phenomena occur during the action of *healthy* nervous arrangements, and constitute, as Hughlings Jackson ³⁷ insisted, a problem in physiology; they represent "physiology in difficulties." It is the more imperative to make this cardinal principle clear because it has been persistently ignored or minimized by many workers, especially in continental schools. To assign tremor and choreo-athetosis to destructive lesions of the corpus striatum is not only impossible, but absurd; a "hole" in the corpus striatum—for that is what outfall of parenchymatous cell-fibre-systems means—cannot cause any movement whatever. Involuntary movements are the derivative of living mechanisms, and *our problem, therefore, is not what mechanisms are paralysed, but what mechanisms are active.*

(2) As an obvious corollary, if after development of tremor, chorea, or athetosis a destructive lesion leads to cessation of involuntary movement, then that lesion must be so situated as to interrupt the neural mechanism that has been producing the movement. Hence the special significance of all cases in which this class of movement has permanently *ceased* after a destructive lesion of one kind or another. Cases of this sort, it so happens, are rare, a fact which renders their careful investigation and recording the more desirable.

(3) A third consideration bears on the question whether the symptoms are produced by excitation or are of the nature of release-phenomena. English neurology, following the lead of Hughlings Jackson, sees many objections to the idea of production of year-long involuntary motor activity by irritation; yet the doctrine that excessive movement is thus caused still has adherents. To determine the exact part played by irritation in the mechanics of symptom-production is no doubt difficult; of the precise nature of the process whereby cell activity passes from a static to a kinetic stage we are still in ignorance. With some show of reason, therefore, it might be held we cannot prove that the liberation of stored-up energy is *not* due to some form of stimulation or excitation; even with withdrawal of inhibition some stimulus might still be postulated to initiate discharge. The argument must be based accordingly on broader grounds.

(a) The duration of some of the cases of involuntary movement we meet with clinically puts irritating lesions out of court. That irritation should last practically a lifetime, as in some instances of Huntington's chorea, is scarcely conceivable, aside from the fact that we are equally at a loss to figure to the mind what sort of

irritation might thus exist and continue indefinitely. It is equally difficult to imagine irritation of nerve-cells giving rise to the chronic tremor of paralysis agitans.

(b) As long ago as 1876 Jackson said, "in some cases it seems to me to be evident that contemporaneously with the loss of function of a centre there is a rise in activity of the next lower centre." This is true not merely of diseased but also of healthy states. The activities of lower centres exhibit themselves freely in the absence of control from higher centres; conversely, the latter may compel the former to suspend their duties temporarily, as is readily exemplified by the action of volition on the respiratory mechanism. A sentence of Hughlings Jackson's,³⁸ of which so far as I am aware no particular notice has hitherto been taken, seems to me to contain a truth of much significance (to which further allusion will be made); the centres of the lower physiological levels "are considerably independent of the directive influence of the higher levels, although much under their negative (inhibitory) influence." By reference to a physiological hierarchy of this kind the phenomena of involuntary movement can be more satisfactorily explained than by any process of irritation.

(4) An integral part of the problem before us bears on clinical distinctions between different kinds of movement. If, as we shall see is the case, fundamental clinical differences can be established between types—for example, between tremor and choreo-athetosis—then the probability of their being mediated by equally different physiological mechanisms becomes a practical certainty. That the same mechanism should be at one time responsible for athetosis, at another for tremor, is unthinkable. The import of this contention will become more obvious as we proceed.

Chorea

In reality a symptom, chorea is the name given to more than one disease. Sydenham's chorea is etiologically distinct from Huntington's chorea; symptomatic chorea is an episodic syndrome arising in the course of or as a sequel to encephalitis, cerebral vascular disease, tumours, and so on. In the two former, and indeed, unless they are exceptional, in the latter group also, the lesions underlying the clinical symptoms are characterized by diffuseness and are ipso facto valueless for the drawing of exact (localizing) deductions therefrom; only in the case of small and circumscribed vascular lesions or tumours is it practicable to

associate the choreic symptom with the pathological finding. Again, in the choreas that bear the names of Sydenham and Huntington symptoms of a different class are frequently in evidence. Instead, then, of endeavouring to localize choreic movements by resort to an uncertain and, for the above reasons, unsuitable pathology, I urge the importance of the reverse procedure ; we shall learn more of the nature and localization of chorea by a precise investigation of its clinical phenomena than in any other way.

Clinical Characters of Chorea

All of us recognize a case of ordinarily well-marked chorea when we see it, difficult though the finding of a satisfactory definition of choreic movement may be. The following are some of the characters of this hyperkinesis.

(1) The spontaneous movements of chorea offer an intimate resemblance to those executed at the bidding of volition. They appear to be as complex (on occasion), as co-ordinated, and as purposeful as those of a frankly voluntary kind. Of their occasional or frequent elaborateness no doubt can be entertained ; as a general rule, the muscular components of the movements take their specific physiological shares in synergia and sequence (to this rule there are important exceptions, as we shall shortly see) ; and each fresh movement appears to be directed to an end—which is never attained. Choreic motility may be described with sufficient accuracy as subjectively purposeful but objectively purposeless ; in appearance only are the movements purposive, but to describe them as purposeless without the qualification just given would be rather misleading.

Many previous writers have emphasized this general resemblance to voluntary movements. Hughlings Jackson (1873) said, " they are not mere spasms and cramps ; they are an aimless [i.e., objectively aimless] profusion of movements of considerable complexity, much nearer the purposive movements of health." Again (1875), " the nature of the movements in chorea (a profusion of real movements—smiles, frowns, and gesticulations) is strong evidence that the part diseased serves in highly special and complex co-ordinations, and thus that it is a part very high up in the nervous system." Cecil Wall ³⁹ (Bradshaw lecture on Chorea, 1920) also says, " I prefer the term ' spontaneous ' to ' involuntary,' not because the latter is incorrect, but because the movements, though occurring independently of the will, are of the same type as movements executed as the result of volition."

(2) A further feature is their brevity and unsustained character, their abruptness, almost explosiveness. They rise and fall, wax and wane, with a peculiar rapidity. Occasionally, and by comparison rarely, a lingering decontraction obtains; the movement comes to an end as if with reluctance. Corroboration of the correctness of clinical observation in this respect is furnished by the electromyographic researches of Stanley Cobb,⁴⁰ who has shown the essential similarity of the action-current curves for voluntary and for choreic movement, the difference residing in the shortness of choreic contractions, as a rule, and in their lack of sustainment.

Now it is of especial significance that *this feature of the spontaneous movements is equally characteristic of much of the patient's voluntary actions*. As a fact, too, long after the involuntary gesticulations, as such, have vanished, the choreic child will often still exhibit noteworthy abruptness of *volitional* movement. The "choreic tongue," which flies back into the mouth at the signal for release, is an example in point. The child shoots his arms out in front when asked to extend them; told to stand up, he rises from his chair with an almost amusing smartness and promptness. Walking down the ward and asked to turn round, he performs the movement with an abruptness that almost unbalances him. That one and the same clinical feature should characterize *both* voluntary and involuntary actions has an important bearing on localization and pathogenesis, as we shall duly see.

(3) A third point in respect of choreic movements is their separate-ness. However rapidly they follow each other, they do not, as a rule, blend into a continuum of motor activity, as is the case with athetosis. They remain discrete, and as long as they so remain, their co-ordination is comparatively undisturbed. To this rule, however, there are exceptions. On occasion, one involuntary movement has not ceased before its successor develops, and when coalescence of this kind occurs the clinical result is a medley of spontaneous muscular activity not to be distinguished in any essential particular from that of athetosis. I had for long under my care an aggravated case of senile chorea (largely right-sided) in an old lady, from whom the records reproduced in Figs. 23, 24, and 25 have been obtained. With the slow-motion camera the *apparently* purposeful and co-ordinated nature of the never-ceasing large and irregular movements is seen; if, however, arm or leg is so fixed as to minimize displacement, it is easy to convince oneself that contraction follows contraction in a confused, blended, and erratic fashion,

and that physiological laws governing action and timing of ordinary muscular components of co-ordinated movement are in complete

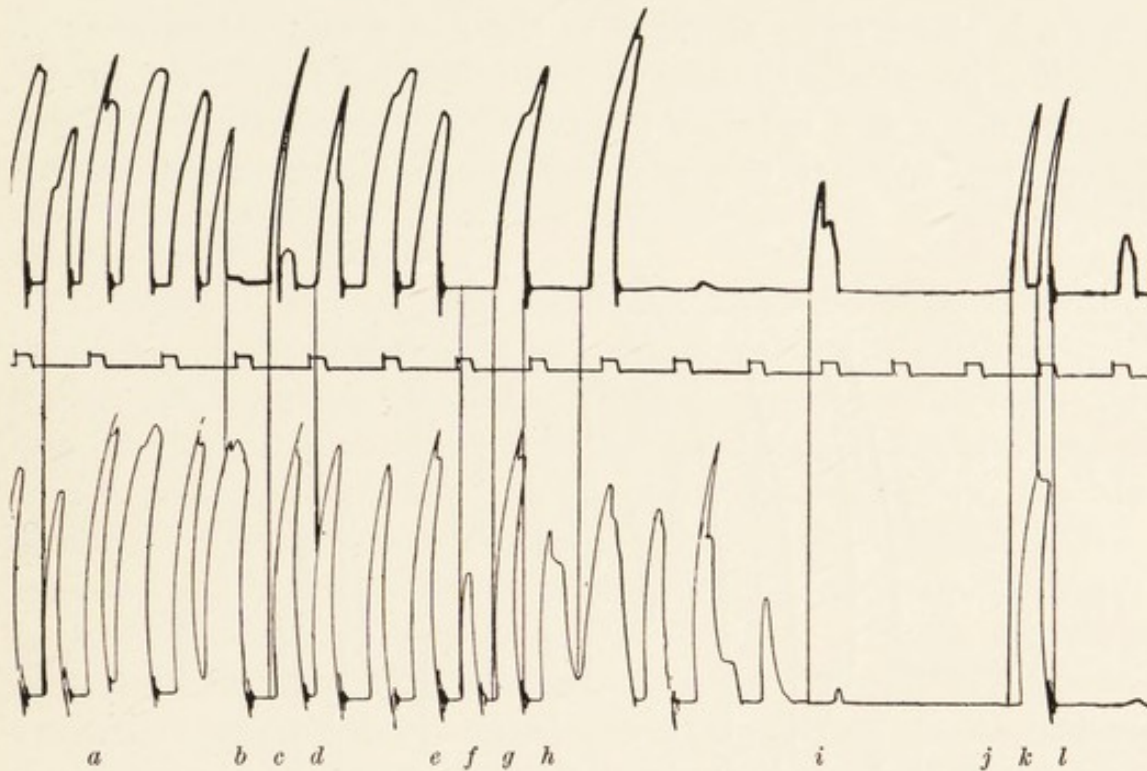


FIG. 23.—Case of senile chorea. Upper tracing gives contractions of quadriceps and lower tracing of external group of hamstrings (R. leg). Simultaneous record.

abeyance. In a word, *Sherrington's law of reciprocal innervation no longer holds, disease having led to its negation.* For instance, atten-

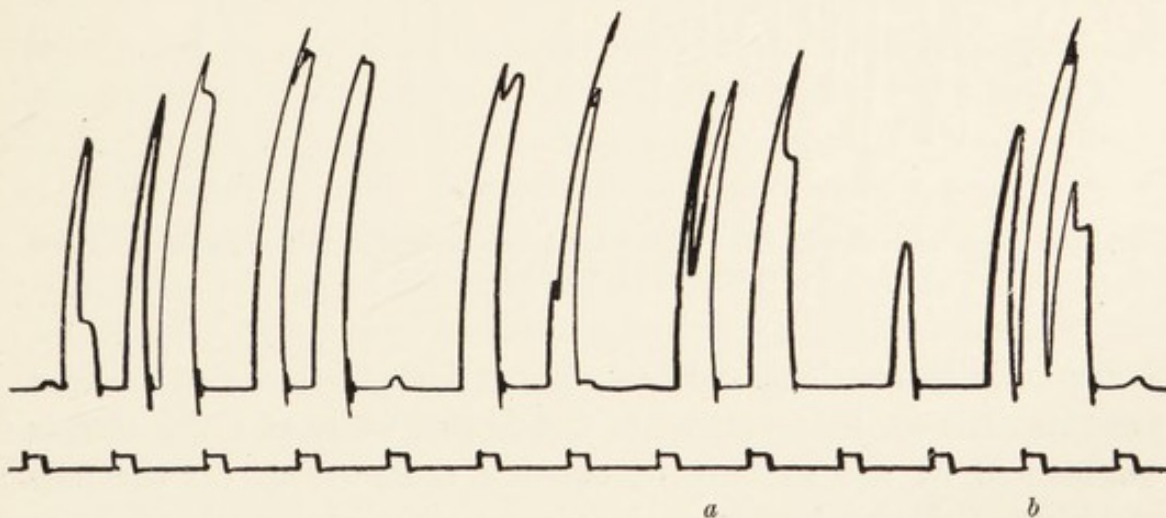


FIG. 24.—Same case as Fig. 23. Contractions of R. biceps.

tion is directed to Fig. 23, which gives a simultaneous tracing of quadriceps (above) and external group of hamstrings (below).

Between *a* and *b* are five contractions of the extensors to four and a half contractions of the flexors, the opposite muscles contracting with practical simultaneity; the extensor contraction immediately after *c* is about twice as rapid as the flexor contraction after *c*, though of practically the same extent; between *f* and *g* are simultaneous flexor and extensor innervations; between *g* and *h* the flexors alone contract, then at *h* both groups contract together, but

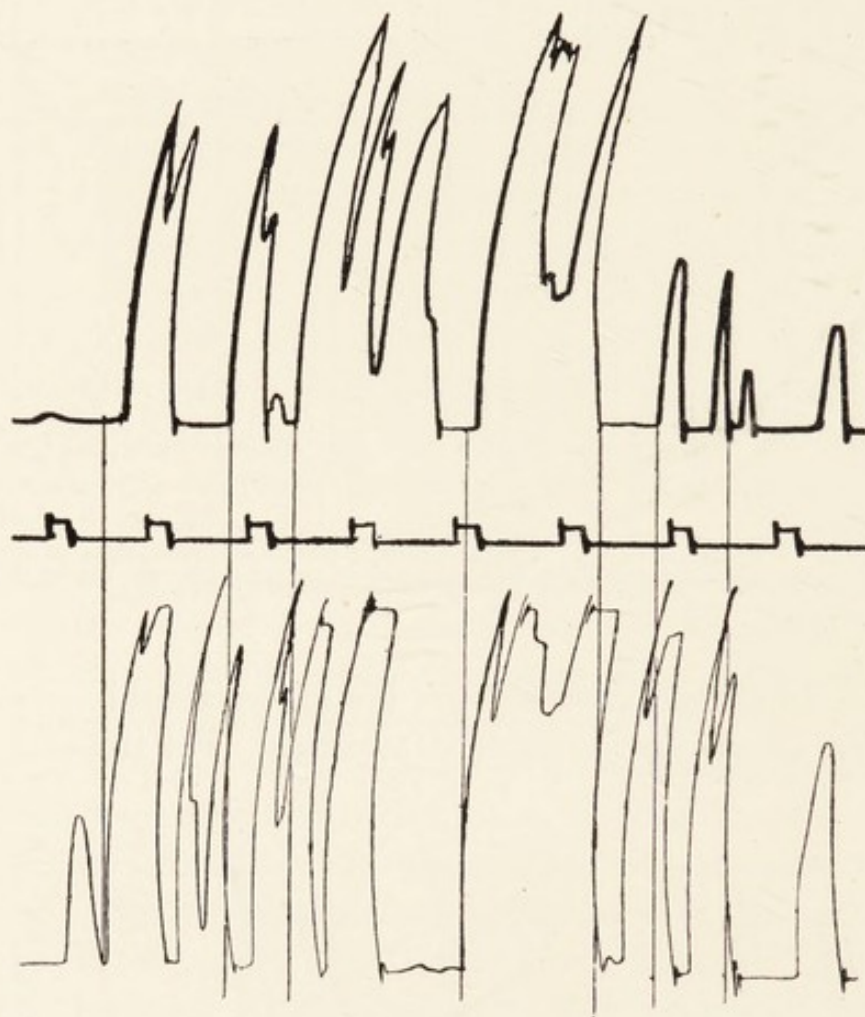


FIG. 25.—Same case as Fig. 23. Simultaneous records of contractions of R. biceps (above) and R. triceps (below).

flexor activity continues while extensor ceases; between *j* and *l* are two extensor innervations to one flexor, while at *k* the former commences at the summit of existing contraction of the antagonists, yet the two end their decontraction together. Thus all kinds of pathological combination of agonist-antagonist activity are exemplified in the record. Fig. 24 is from a tracing of the right biceps of the same case, and shows not only complete irregularity in time and extent of the individual contractions of the muscle, but

also (at *a*) how a second contraction supervenes on a first, before the latter has ceased. At *b*, again, three contractions follow on each other without a single break. In Fig. 25 are seen the contractions of right biceps and triceps of the same case, and by comparing these as they occur between the various ordinates drawn on the record dissolution of physiological "give-and-take" can at once be recognized. No clearer demonstration of the *complete interruption of normal physiological reciprocal innervation during confluent choreic activity* could be found.

(4) This interference with normal components of a movement producing joint displacement is not, as a rule, to be discovered except in what I call "confluent chorea." Years ago, however, the late Dr. Beevor drew attention to the occasional occurrence in acute chorea of what he termed "paradoxical contraction of the abdomen." Normally the diaphragm descends during deep inspiration and at the same time the recti abdominis slacken; the upper part of the abdomen is seen to come forward, or, if the subject is recumbent, to rise, with descent of viscera associated with diaphragmatic contraction. Now sighing inspiratory movements are fairly frequent in severe choreas, and on occasion, with inspiration, the abdominal muscles can be clearly observed to contract paradoxically, rendering the upper abdomen concave instead of, as normally, convex. Here is a definite clinical instance of lack of harmony between the constituent muscular units of a choreic movement, and of abrogation of physiological sequence.

I have studied several cases of Huntington's chorea from the standpoint of normal muscular action, using for the purpose the scheme previously described. Interruption of the normal interplay of constituent muscles is but rarely found, though when it occurs it is definite enough. For example, I have seen dissociation of normal elevation of the eyebrows with upward deviation of the eyes, also of head extension in depression of lower jaw against resistance. With the patient recumbent, I have seen disturbance of the normal sequence of sternomastoid, rectus abdominis, and rectus femoris action in the movement of rising to a sitting position.

In both voluntary and involuntary movement, therefore, in the case of chorea, proof of the not infrequent interruption of physiological law in the performance of movements requiring harmonious co-ordination of prime movers, synergists, and antagonists, as well as in those which I have called movements of sequence, has been obtained.

The "Choreic Hand"

(5) The earliest mention I have been able to find of the "choreic hand" (though not with this specific term applied to it) is by Francis Warner⁴¹ (1885). It is constituted by flexion at wrist and hyperextension at metacarpo-phalangeal joints, the phalanges being straight or moderately overextended (or very slightly flexed), the fingers usually rather separated, the thumb overextended and

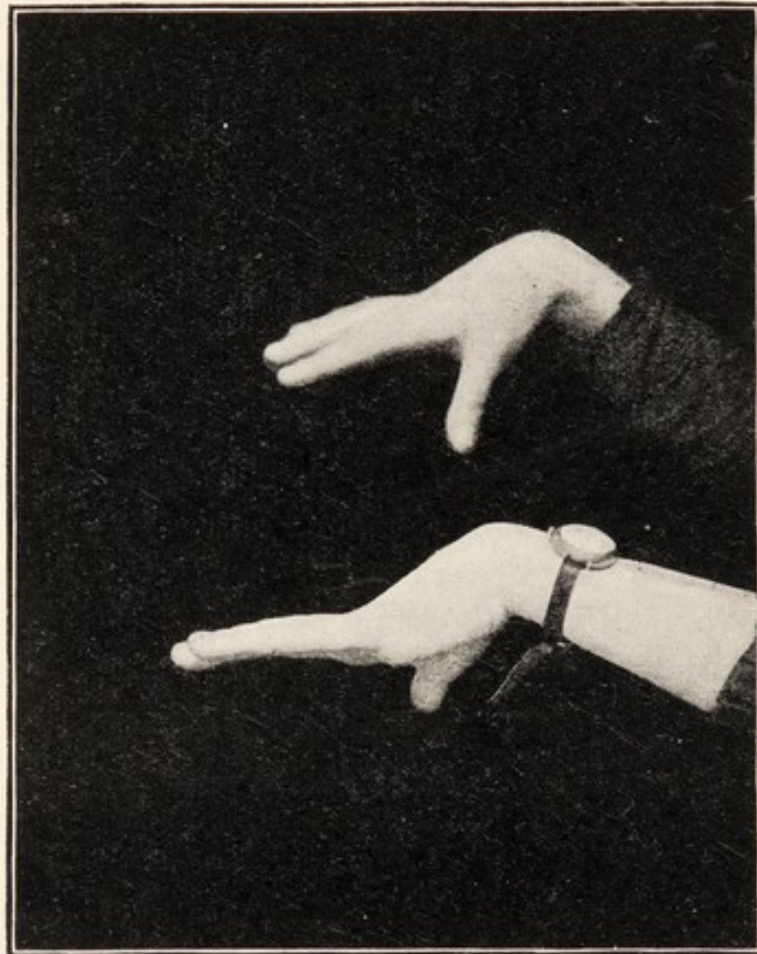


FIG. 26.—The Choreic Hand.

abducted, and often "dipping" (Fig. 26). Warner found this attitude (to which he gave the name of the "nervous hand") inter alios among "those convalescent from chorea," and he also stated: "It is also often seen on one side only, especially in children convalescent from hemichorea." I have for years given attention to the choreic hand and have remarked its presence in an extremely high percentage of cases; it may persist, like the abruptness of voluntary movement already alluded to, long after spontaneous choreic movement has ceased. Without exception it will be found

to be more pronounced on the side (if there be such) on which spontaneous movements are (or have been) more frequent. Its occurrence, therefore, must be linked with the mechanism or mechanisms implicated in the mediation of these movements.

Now while the posture of the choreic hand may be adopted momentarily in the course of a spontaneous choreic movement, this is undoubtedly somewhat rare; to demonstrate it most easily the patient is asked to extend his arms in front. The hands then assume the posture of wrist flexion and metacarpo-phalangeal extension, often in a strikingly exaggerated form. When a normal subject is requested to hold out his hands a difference is usually noticeable according as the movement is made energetically or gently; in the latter case they are seen to fall into a posture of moderate extension at wrist with probably slight flexion of fingers; in the former the fingers will be rather more extended, though the wrists will probably be extended, as before. Adoption of this attitude by the patient, however, is by no means merely an expression of energetic abruptness of voluntary movement, for it is readily obtained in numerous instances where the movement is not so characterized.

The choreic hand, as a moment's consideration shows, is the "corresponding opposite" of that posture of the hand seen in everyone as the arms depend in rest by the side, or, for that matter, in any easy resting position—viz., extension of wrist and flexion of fingers—the expression of activity of the normal synergic muscles for grasping. The wrist extensors are the normal synergists for the antagonistic long finger flexors and their physiological association is intimate and constant. Now, as anyone can discover for himself, it actually requires a conscious effort to assume the choreic attitude voluntarily, and the more pronounced the attitude the more is he aware of strain and effort in making and maintaining it. One reason, doubtless, for this feeling resides in the infrequency, indeed, the rarity, of any voluntary movement or posture of this particular sort. Yet this is the precise posture effected by a disordered condition of the neural mechanisms concerned. That is to say, in the case of the choreic hand there is an "involuntary" (for though occurring in the course of a "voluntary" movement it is certainly not assumed consciously by the patient) adoption of a posture the opposite of that constantly seen in normal persons in a resting state, and of that ordinarily found in the hand when the arm is held out; in other words, a "*physiological*

shift” occurs from one posture to its corresponding opposite. When the sole of the foot is stimulated a normal movement of plantar flexion takes place ; in states of abnormal function of the pyramidal system a similar cutaneous stimulus, applied in a similar place, gives rise to a movement of plantar extension—there is a physiological shift from one *reflex movement* to its opposite. This perhaps provides us with an indirect analogy by which to endeavour to explain the choreic phenomenon ; it may be taken to suggest a postural change associated with abnormal function of the corticospinal system, though this abnormal state need not originate on the motor or effector side.

The Pronator Sign

(6) Other “little signs” in chorea are indicative of a condition of disordered corticospinal function. In unilateral instances Babinski’s “rising sign,” described by him for cases of hemiplegia, is often obtainable on the affected side. Occasionally an extensor plantar response is found in chorea ; I have seen it more than once but do not stress the point unduly. Much more frequent is the “pronator sign,” the arm assuming the same position of pronation or semi-pronation at the elbow as is seen in hemiplegia. When the choreic holds out his arms the thumbs are often seen to “dip” more than normally, indicative of some forearm pronation ; a better way is to have the patient extend his arms above his head, when the palms are observed to turn outwards, sometimes to a pronounced degree, thus revealing an involuntary over-pronation at the elbow (Fig. 27). In a paper⁴² on decerebrate rigidity in man allusion has already been made to some of these phenomena and the suggestion put forward that choreic postures—the choreic hand, the pronator sign—are pro tanto indicative of the partial release from voluntary control of mechanisms that are wholly released in complete decerebrate rigidity. The suggestion has been misunderstood by some subsequent writers, but from what has been outlined above it will be seen to indicate simply the fact that certain analogies obtain between the results of release of function in chorea and of release in disorder of the corticospinal system. In another Chapter (see page 138) I have argued from clinical data that the results of complete single release (hemiplegia, paraplegia) are not identical with those of complete double release (decerebrate rigidity, cortical + striatal efferent systems).

Paralytic Chorea

As further evidence of the point at present under consideration, reference may be made to cases of chorea signalized rather by weakness and inactivity than by spontaneous movements. The complaint in these cases is always one of losing the use of the limb. Objects are dropped from the hand; the arm hangs practically motionless; weakness of grasp is often pronounced. Such cases



FIG. 27.—The Pronator Sign.

are by no means rare, and were described originally as "paralytic chorea" by Sir William Gowers⁴³ in 1880, and as chorea mollis by Samuel West. In any ordinary case, too, some muscular weakness can usually be discovered. In my experience of flaccid chorea, the greater the weakness the fewer are the spontaneous movements; with return of power the latter may for a time increase; hence the apparent paradox that the child seems to the parents to be "getting worse" when as a fact improvement has set in.

The significance of such cases will be better understood if we bear

in mind the important principle that "weakness" or hypokinesia and "involuntary movements" or hyperkinesia are but different aspects of activity of the same mechanisms; in one case there is underaction, in the other, overaction, of motor function. Hypokinetic chorea is as it were the obverse of hyperkinetic chorea; and we should not, therefore, seek a *different* mechanism for each, but rather expect to find that in the case of the latter the efferent corticospinal system is less disordered than in the former.

(7) The general state of muscle tone in chorea is of significance in relation to problems of localization and pathogenesis. Commonly speaking, choreic musculature is definitely hypotonic, as regards both the usual and the paralytic type, and allows of greater extent of passive movement than in normal persons of the choreic age, say 7 to 15. Further, in numerous instances of Sydenham's chorea the knee-jerk exhibits the pendular character found in hypotonic cases of cerebellar origin. Though the hamstrings are passively stretched by the shortening reaction of the quadriceps their tone is relatively so low that a "stretching-contraction" does not take place; the leg, therefore, swings like a pendulum. The knee-jerk differs not in kind but in degree from the cerebellar knee-jerk, which exhibits the phenomenon often in a more striking fashion.

(8) One other clinical consideration remains before we seek to draw conclusions as to pathogenesis from our clinical data. Among the spontaneous movements of chorea are to be found many involving the muscles of facial expression; across the choreic's face flits an exaggerated smile, to be followed in a moment by an expression of preternatural gravity. Frowns, smiles, sighs, chase each other unendingly; when the choreic tongue flies back at a word into the mouth the face is often liable just then to be disturbed by a series of expressional activities. These transient emotional or mimic displays, however, are essentially *movements*; one does not, as a rule, observe individual muscular twitches, myoclonic contractions of individual muscles, never any localized muscular caricaturing as in facial spasm. If unilateral grimacing occurs, it offers the same features as facial tic. Almost invariably those movements are bilateral which in health are bilateral; dissociation occurs in lower facial movements but not in upper, consonant with normal physiological law. I have not observed unilateral contractions of the frontalis, for example. We must not imagine the fragments of emotional facial expression exhibited haphazardly by the choreic to correspond to any existing emotional state of the

moment ; rather are they to be regarded as a release of the efferent side of the mechanism for the expression of the emotions. In another Chapter (see page 284) evidence has been adduced suggesting a cortical (non-rolandic) site for the central representation of mimic expression, and if this hypothesis is worthy of credence we may suppose in chorea a double condition of release : (1) The "voluntary" motor system (corticospinal system) is usurped by "involuntary" movements ; (2) a normally "involuntary" efferent motor system (that for the expression of the emotions) is the seat of movements not occasioned by normal sensory (emotional) stimuli, but by abnormal (choreic) stimuli. Previous allusion has been made to the possibility of such a double release.

Summary of Clinical Analysis of Choreic Symptoms

Summarizing this analysis of choreiform phenomena, we have noted the intimate resemblance of choreic movements to voluntary movements, the existence of "little signs" pointing to disturbance of corticospinal function, the occurrence in both voluntary and involuntary choreic action of identical clinical characteristics ; we have seen the connexion between paralytic and hyperkinetic chorea, and realized how these are but differing aspects of derangement of the same efferent mechanism ; while the occurrence in confluent chorea (as I call it) of motor inco-ordination which clearly demonstrates abrogation of the law of reciprocal innervation by disease has also been exemplified.

Now the argument from these clinical data which is here offered may be stated as follows : Complete paralysis, as in, say, anterior poliomyelitis, is never under any circumstances associated with the appearance of involuntary movements in the affected limbs ; similarly, in the case of complete flaccid hemiplegia no involuntary movement (post-hemiplegic chorea or athetosis) makes its appearance until there is clinical evidence of some return of power in the paralysed limb or limbs. Even in cases of severe chorea the corticospinal system is never paralysed, but the greater the degree of paralytic chorea the fewer are the spontaneous choreic movements. It is difficult to resist the conclusion that *choreic motor disorder manifests itself through the pyramidal (corticospinal) tracts, which must be in a condition of relative integrity for hyperkinetic symptoms to appear, and yet exhibit some defect of function as indicated by the above-mentioned minor signs.* Hence the voluntary movements of choreic patients have some of the characters of the

involuntary movements, as we have seen ; the latter take the same route and usurp it, at the expense of the former. At the same time, choreic motor derangement need not actually originate on the motor (effector) side of cortical mechanisms, as we shall see.

Further argument may conveniently be postponed till the clinical phenomena of athetosis have been subjected to analysis.

Athetosis

As with chorea, so with athetosis, a typical case is easy of recognition, though to define the symptom is not equally simple. In 1871 Hammond, who first drew attention to it, stated that "it is mainly characterized by an inability to retain the fingers and toes in any position in which they may be placed, and by their continual motion," and in another passage he said that "the movements are slow, apparently determinate, systematic, and uniform," in contrast to those of (post-hemiplegic) chorea, which are "irregular, jerking, variable, and quick." The contrast between the two, however, is by no means so definite as this description might lead one to suppose.

Clinical Characters

(1) Athetoid movements are characterized by relative slowness, in apparent contrast with the relative rapidity of choreic movements, and by the perpetual blending of one movement with another, leading to the "continual motion" of Hammond. They form a mixture of irregularly synchronous contractions of opposite muscular groups, so that on occasion the limb or a segment of it is immobilized momentarily in an attitude. The "mobile spasm" flows indifferently into muscular units, the result being a medley of contraction entangling the limb, as it were, and leading to no accomplishment.

During this irregular muscular display varying rigidity naturally makes its appearance, yet unless the athetosis is associated with some degree of paralysis there is little, if any, actual fixed rigidity, and this generally bears some proportion to the amount of paralysis. In fact, during temporary relaxation of athetotic activity the limb musculature is frequently, if not constantly, hypotonic. Voluntary innervation of the affected part is always laborious and commonly aggravates the athetoid contractions, at least within a very short period ; voluntary control over them is fleeting and imperfect.

(2) Closer analysis can be made conveniently by means of double tambour tracings, care being taken by adequate limb fixation

to ensure the absence from the record of fortuitous (non-athetoid) movements. Fig. 28 reproduces a tracing taken from the left forearm in a case of adult athetosis (of vascular origin, and progressive); the upper tracing represents the contractions of the extensor communis digitorum, the lower, of the flexor longus digitorum. At *a* practically synchronous contractions of these opposite muscles occur; at *b* a series of four similar concomitant contractions is seen; between *c* and *d* flexor activity is more pronounced than extensor; at *d* the extensor group contracts vigorously during flexor contraction; and at *e* flexor contractions take place by

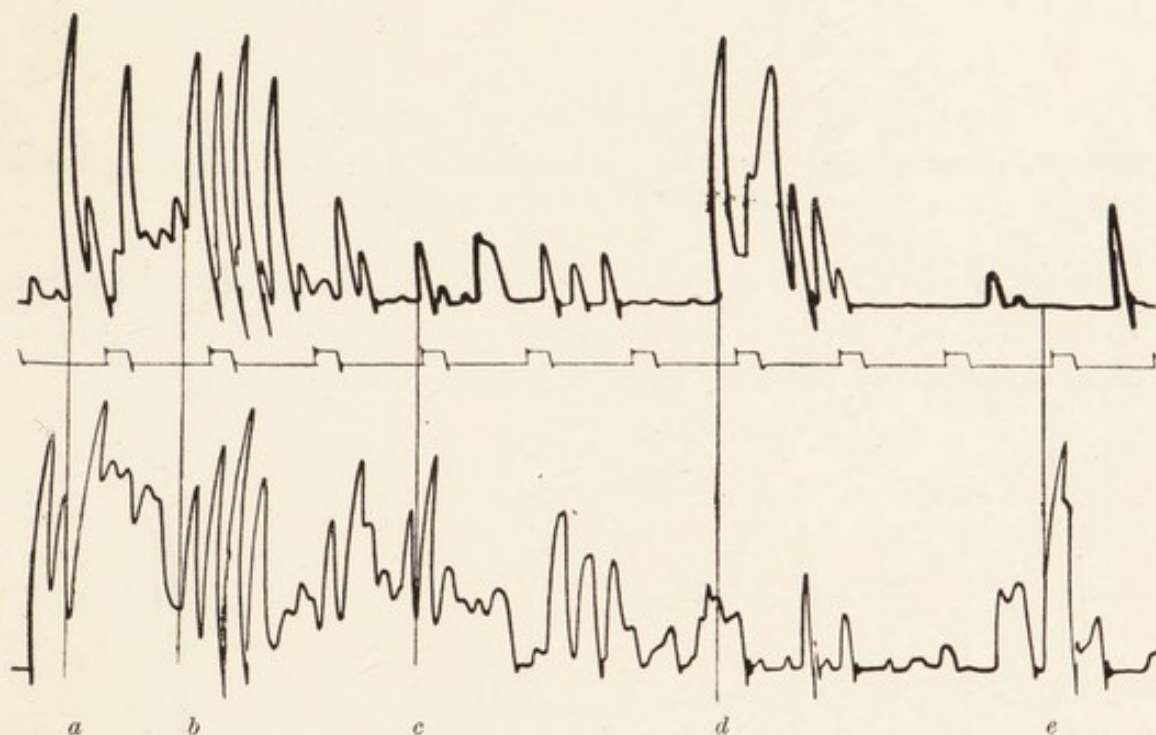


FIG. 28.—Athetosis. Upper record, contractions of left extensor communis digitorum: lower, left flexor longus digitorum.

themselves. Irregularity of individual contractions is well-shown in Fig. 29 (from the same case), recording the innervation of the long finger flexors during a period of extra-excitability. Here contraction follows contraction in an extremely confused fashion, one being often thrown in before the muscle has relaxed from the previous contraction. Fig. 30 records simultaneously the contractions of the biceps brachii and triceps of the same case; it shows, between *a* and *c*, three contractions of the biceps to one (tripled) of the triceps; at *b* reciprocal innervation is completely in default; at *c* flexor and extensor contractions are almost but not quite synchronous; at *d* the triceps contracts and a little later the biceps,

both relaxing together ; and at *e* the former contracts in four quick innervations by itself.

Another series of records have been taken from a case of athetosis

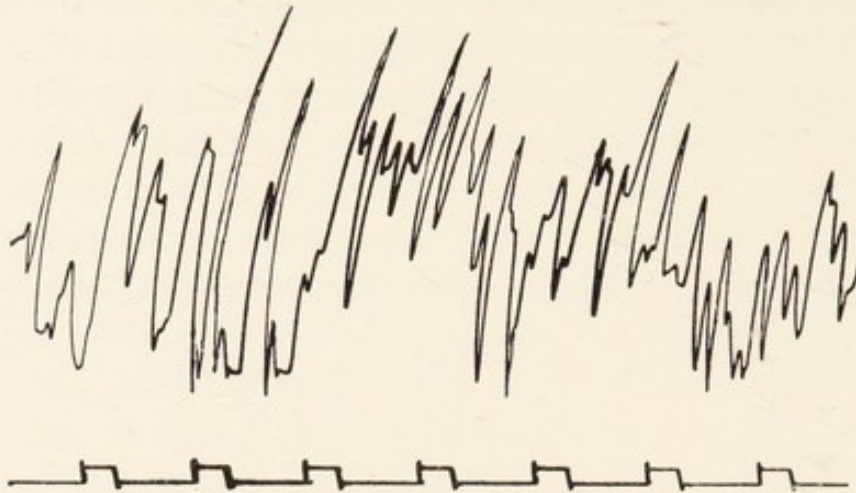


FIG. 29.—Same case as Fig. 28. Contractions of left flexor longus digitorum during a period of extra-excitability.

in which I have made use of the extensors of the wrist and fingers. Normally, the former should relax (after synergic contraction during closure of the fist) when the fingers are extended.

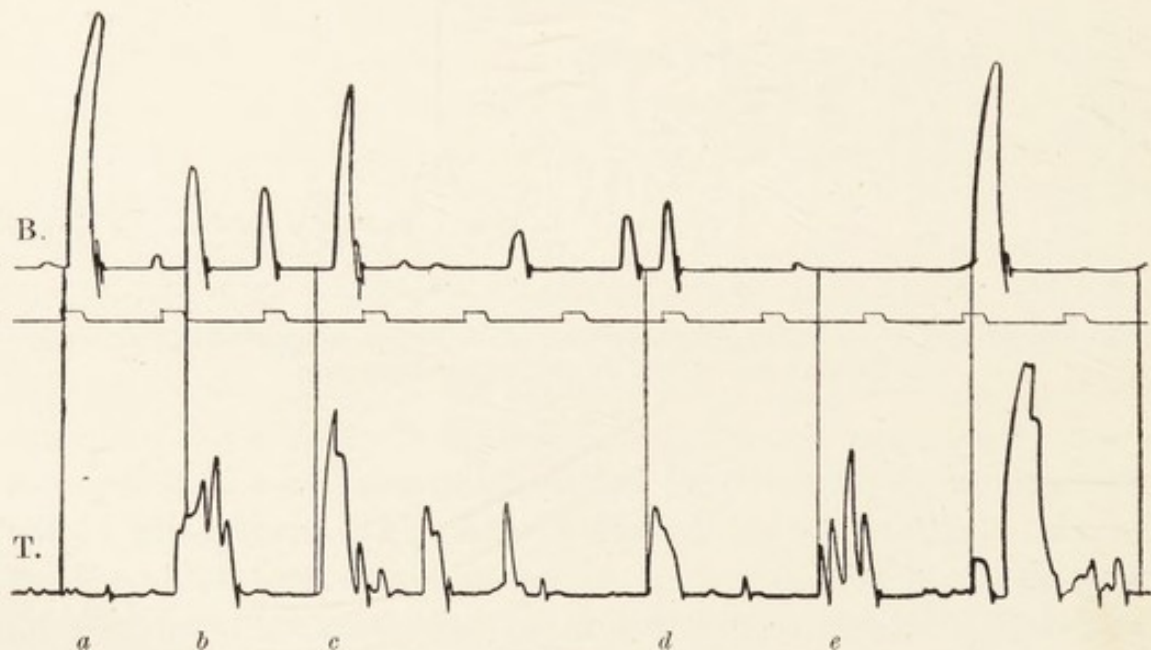


FIG. 30.—Same case. Contractions of biceps and triceps.

In Figs. 31 and 32 are represented two different results obtained from this patient ; in Fig. 31 wrist extensors decontract appreciably *before* the contraction of finger extensors ; in Fig. 32 finger extensors contract normally, yet wrist extensors hold their contraction for a time, then at length relax, but contract once more. Such physio-

logical modifications are beyond the power of volition to effect. Nothing could more conclusively demonstrate the *complete abrogation by disease of the normal physiological law of reciprocal innervation, and this is characteristic of athetosis*; in paralysis agitans, as we have seen, it is not annulled.

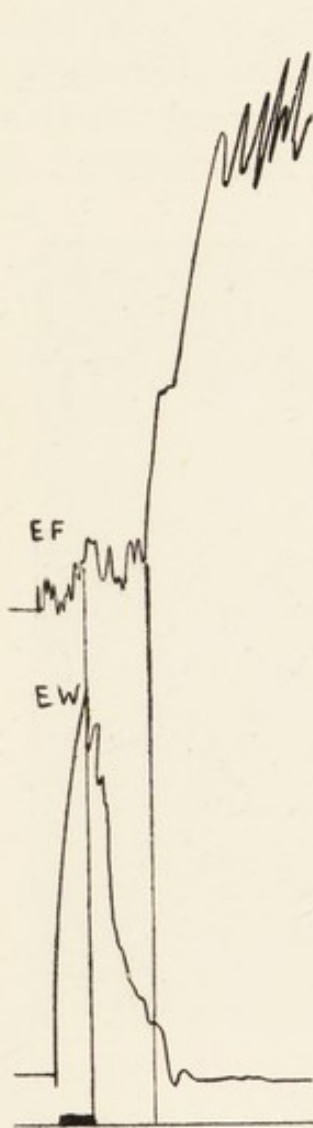


FIG. 31.—Athetosis. Decontraction of wrist extensors before finger extensors contract.

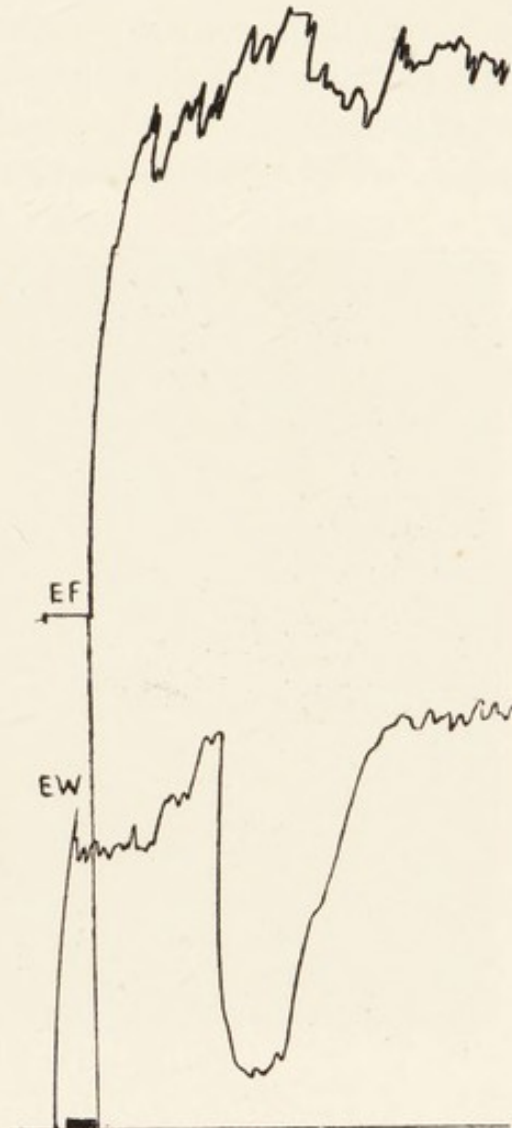


FIG. 32.—Same case as Fig. 31. Wrist extensors fail at first to decontract when finger extensors contract: after later relaxation they contract again.

(3) Owing to the essential variability of athetotic contraction no enduring posture might perhaps be expected, yet the "athetoid hand" is sufficiently definite to merit attention (Fig. 33). A saddle-shaped posture of the fingers is common, the terminal phalanx being in flexion while the proximal phalanges are in extension; with this extension (often hyperextension) at metacarpo-

phalangeal joints is coupled a notable flexion at wrist, so that the hand often assumes that attitude which we have seen is of extreme frequency in chorea and which, as we have also seen, represents a physiological shift from the posture of the normal hand at rest. In athetosis, further, a tendency for pronation or hyperpronation of the forearm (the "pronator sign") is equally common.

(4) Comparing, now, the symptoms of chorea and athetosis, we find the resemblances more impressive than the differences. In respect of the latter, we have seen that choreic movement is discrete and rapid, while athetoid movement is slow and confluent. But

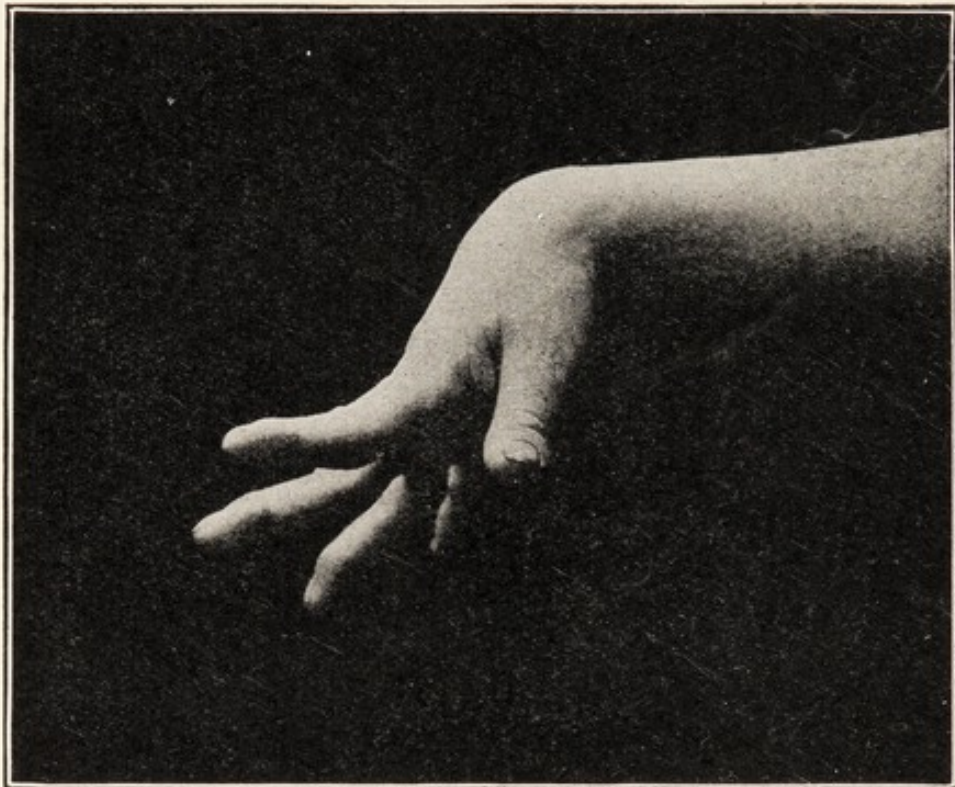


FIG. 33.—The athetoid hand.

this difference is more apparent than real; athetoid action is slow largely because it is confluent, and, on the other hand, confluent chorea results in slowing of the spontaneous movements. Besides, in some athetoses a certain degree of discreteness and quickness is observable. In respect of resemblances, in each the type of motor derangement is complex, elaborate, and specialized; changeableness of movement is a prominent trait (i.e., absence of essential repetitiveness); the law of reciprocal innervation is at fault in athetosis and in confluent chorea equally; the movements, "caricatures" though they are in athetosis more than in chorea, have the appearance none the less in the case of the former of being

subjectively purposeful—as for grasping or relinquishing an object, etc.—though they are objectively purposeless. In both, a degree of hypotonia is commonly discoverable; resemblances between the athetoid and the choreic hand are sufficiently obvious; involvement of the mechanism for emotional expression, if found in athetosis, is commonly bilateral, as in chorea, though to this there are exceptions. It is difficult to gauge the amount, if any, of actual weakness of the musculature in athetotic cases, since (*a*) they are not infrequently accompanied by definite corticospinal derangement (hemiplegia or hemiparesis), from which it is desirable to attempt to separate the athetotic element; (*b*) volitional innervation is constantly impeded by athetoid activity. Enough, however, has been said to justify fully the common grouping of choreic and athetotic movement under the general heading of “choreo-athetosis.”

Relation of Athetosis to Tonic Innervation

(5) The condition described as “tonic innervation” is constituted by inability to relax voluntarily a given volitional innervation. As the patient voluntarily contracts his flexors (say) decontraction of antagonists (extensors) takes place, according to the law of reciprocal innervation; when next he would reverse this movement, he is able voluntarily to innervate the extensors, but no corresponding inhibition of the flexors takes place; these remain contracted, it may be for many seconds. From what we have been able to show by clinical methods, a condition resembling tonic innervation, in fact practically identical with it, may occur in athetosis. I have a number of observations confirming this statement, which, for that matter, was fully exemplified in a paper⁴⁴ on “The Phenomenon of Tonic Innervation.” The symptom was then described as occurring in cerebral cases (frontal) unassociated with athetosis, and as being seen mainly if not solely during volitional movement. In the case of athetosis it is found, when present, during involuntary activity, in the sense that antagonists may remain definitely contracted though the opposite muscles are rising into contraction, but it often also develops in typical guise during voluntary movement. I have already commented on the fact that no phenomenon of this kind has come under my notice in any of the scores of Parkinsonian cases personally investigated.

Clearly, it can only be explained satisfactorily by regarding it as a derangement in the proper relation between impulse and

inhibition—i.e., as a disorder of reciprocal innervation. Thus our clinical study of athetosis leads us again to the view that *its essential and underlying basis is precisely this interference with normal laws governing physiological interaction of component muscular units in movement, and it is immaterial whether this be voluntary or involuntary*, for no discriminating line can be drawn between the two.

Pathogenesis of Choro-athetosis

On the clinical side, evidence has been adduced indicating, both for chorea and for athetosis, that the following considerations bear on the problem of pathogenesis.

(1) Since the movements are active and continuing, for their existence relative integrity of some efferent path is a *sine qua non*. Many cogent arguments suggest that this path is the corticospinal path. I may refer the reader, for a moment, to the summary of the clinical analysis given at the close of the sections on chorea, and supplement the deductions there drawn by one or two more matters bearing on the topic.

Many cases of infantile cerebral hemiplegia with Jacksonian fits are accompanied by athetosis. This association is very common and in itself highly suggestive, for it proves conclusively that a degree of impairment of pyramidal function is not inconsistent with the appearance of athetoid movement. We assign the hemiplegia and the epilepsy to encephalitis of the cortex and implication of the corticospinal tract, and it is reasonable to assume in *these* cases a *cortical origin for the athetosis* also. Take a single illustrative case.

A girl of 14 had suffered from infantile cerebral hemiplegia with athetosis and Jacksonian epilepsy since she was one year old. The aura to the fits was the appearance of "twinkling red, blue, and white lights" in the right field, followed by twitching in the right face and right fingers, which spread in typical Jacksonian fashion. For twenty minutes after each fit all athetosis disappeared from the right limbs.

On the sensory side, there was complete astereognosis in the right hand, with only slight diminution in the appreciation of tactile stimuli.

Apparently, an exhaustion paresis of the right corticospinal system made the athetosis temporarily impossible; only with recovery of power did it begin again. The evidence in this case pointed unmistakably to a cortical origin for all the symptoms.

I refer next to the case recorded by the late Sir Victor Horsley⁴⁵ in his Linacre Lecture of 1909. A youth of 15 had had left hemi-

plegia with severe athetosis since an attack of scarlatina at the age of seven. Horsley found at operation that the right postcentral gyrus was reduced to half its normal size—a very significant observation. He excised the cortical motor area for the left arm, and the athetosis thereafter vanished entirely and had not returned when the patient disappeared from notice some fourteen months later. On the other hand, the arm paralysis following the cortical excision had by that time been greatly mitigated. Similar cases have been put on record by Anschütz⁴⁶ and others. Reference, too, may be made to Case XVIII. of Jakob's monograph,⁴ where left-sided athetosis disappeared after a sudden left flaccid hemiplegia. The patient lived for eighteen months after, but the athetosis never returned. (I venture to differ from Jakob as regards the interpretation put by him on the pathological findings in this valuable case.)

Strong additional evidence is hereby furnished of the significance of *relative* integrity of the pyramidal system for chorea and for athetosis. I have never yet found a case of either in which spontaneous movements occurred in the presence of absolute corticospinal paralysis. No movements resembling either have been seen in my clinical cases of decerebrate rigidity in man, and Magnus,⁴⁷ similarly, has commented on their absence in his experimental animals (of various species) submitted to decerebration.

(2) Another line of clinical evidence points to the possibility of implication in choreo-athetotic cases of afferent paths to the cerebral cortex—viz., from cerebellum by regio subthalamica to optic thalamus, whence there is a wide distribution of fibres to the pallium.

The frequent appearance of muscular hypotonia both in chorea and in athetosis has been mentioned above, and the element of inco-ordination is prominent in the clinical picture in both, especially, as concerns chorea, in the case of the confluent variety. In numerous instances of chorea the knee-jerk exhibits that "pendular" quality which pertains largely to cases in which cerebellar function is disordered. Where for other reasons a cerebellifugal (cerebello-cerebral) lesion is likely, we may expect to find some kind of cerebellar symptoms. If, then, other clinical facts argue a cortical origin for a symptom-complex, the coexistence with it of certain cerebellar symptoms can justifiably be set down to the development of a cerebello-cerebral defect of function.

Elsewhere I have on more than one occasion laid stress on

the many clinicopathological cases which point to lesions in one or other division of the cerebello-mesencephalo-thalamo-cortical apparatus being responsible for the appearance clinically of choreo-athetosis. The reality of "Bindearmchorea" (superior cerebellar peduncle, especially in the neighbourhood of Wernekinck's commissure) is definite enough. In several cases of acute thalamic lesion (neothalamus) I have seen pronounced involuntary movements of the choreo-athetotic variety, and in a case of *athétose double* I have found bilateral lesions of the neothalamus (posterior inferior region). A case under my care exhibits the thalamic syndrome with athetosis of the same limbs (arm in particular). I may also cite recent experimental work favouring the view here maintained (Lafora⁴⁸)—though his results require confirmation—and the older and seemingly forgotten experiments of Bickel and Jakob,⁴⁹ who produced "cerebral sensory ataxia" by extirpation of the postcentral cortex, and who specially remarked "the peculiarly explosive character of voluntary movements" after the lesions. This, needless to say, is in fact a choreic character, as has been already discussed. The reduction in size of the post-central gyrus in Horsley's case of athetosis is specially significant, and is paralleled by what I have found in a case of senile chorea (mentioned clinically on page 212). I do not rely merely on individual cases and experiments in support of my general contention, which is based on a series of diverse considerations, of which possibly the intrinsic quality of the movements is the most important.

On a variety of grounds, therefore, the evidence here adduced pleads in favour of the hypothesis offered, that athetosis and chorea are exteriorized via the corticospinal paths, and that behind their appearance is an afferent disorder of regulation, producing choreiform and athetoid characters in movement, ataxia or inco-ordination and hypotonia. This afferent disorder or derangement of regulation is attributable to lesions of the cerebello-mesencephalo-thalamo-cortical system already described. As a consequence, voluntary movements via the corticospinal tracts may exhibit characters corresponding to the functional defects; and these tracts are, continuously or intermittently, usurped also by spontaneous movements of choreiform or athetoid character, movements in many ways closely resembling voluntary movements, over which the patient's inhibitory control remains desultory and incomplete. A persistent or intermittent stream of stimuli keeps up the flow of involuntary

movement; only in sleep are these excitations damped down. During waking life—it may be for years, as we have seen—the stream flows to the cortex and the cortex ceaselessly responds. I do not say that this cerebello-cerebral system exercises normally a directly inhibitory effect* on cortical motor centres, as though it were on a higher physiological level, and that owing to destructive lesions of any part of it the corticospinal system is released into involuntary activity; the origin of the stimuli-producing movement is ultimately sensory—in the cases under discussion, intrinsic stimuli, derived from low or high sensory levels. The route taken by such is presumably that of afferent sensory systems. But defect of cerebello-cerebral regulation—absence of some cerebello-cerebral component in motility—appears to allow or to contribute to overaction of these afferents on cortical efferents; this component should in normal circumstances come into action where afferent impulses are transformed to efferent excitations at cortical junctions, and its absence facilitates their passage. As a consequence, too, the resultant movements exhibit precisely those defects of co-ordination, of reciprocal innervation, of dystonia and “explosiveness,” already analysed. Thus, in a definite sense, choreo-athetoid activity is the activity of cortical reflex arcs; the movements have cortical quality and yet they are involuntary.

Over this mechanism transcortical (“voluntary”) inhibition has very imperfect control. It is a particular case of the general problem presented by the question of *voluntary control over an involuntarily decontrolled mechanism* (see page 250). If the views here offered can be reasonably entertained, they lead to the conception of the possibility of *involuntary cortical motor activity*—a natural development of the hypothesis I have suggested. As far at least as chorea is concerned, the conception appears to me to have much to justify it, and little less in respect of athetosis. *The spontaneous activity of choreo-athetosis, thus conceived, is nought else than a succession of cortical reflexes, high-grade movements largely comparable to those called voluntary except that the patient's volition neither initiates nor inhibits them.*

Further explanation, however, is requisite, in more than one respect.

(1) When I speak of cerebello-mesencephalo-thalamo-cortical

* The fact that recent work, alluded to elsewhere (page 127), attributes to the cerebellum an *inhibitory* function may not be without significance in this connexion.

dysfunction being an integral factor in the appearance of choreo-athetotic movements exteriorized via the pyramidal system, I do not limit the association to an anatomical link in the postcentral cortex. Thalamocortical fibre-systems are widespread, and a frontal radiation is just as definite as any to the postcentral gyrus and no less important. For, on the efferent side, when I have been speaking in various preceding sections of the corticospinal system being usurped by movements of a spontaneous or involuntary kind, I do not wish it to be supposed that I regard the *anatomical* Betz-cell as the *physiological* terminus a quo of efferent motor activity. On the contrary, many lines of evidence are against such a supposition. From the excitable motor area (Betz-cell area) one does not obtain movements of so elaborate or intricate a pattern as are seen in everyday volitional action; the combinations of movement elicitable by the stimulating electrode are anatomical rather than physiological; the march of the movements in Jacksonian epilepsy is not paralleled in normal volitional activity. For the complex movements of chorea, to take an example, we must postulate a motor centre higher in a physiological sense than that of the rolandic motor region, and both clinical, pathological, and experimental evidence suggests that such a centre lies in front of the rolandic area. Relatively short transcortical neural systems link the frontal to the Betz-cell area, and I imagine, consequently, that the afferent disorder of which I have spoken produces its effect by radiation from the thalamus on this postulated frontal motor centre, quite possibly from parietal to frontal cortex, and so on the (anatomical) origin of the corticospinal tracts.

(2) The inability of choreic or athetotic patient to control adequately the abnormal activity of the cortical arcs of which I have spoken can only mean, translated into physiological terms, *impairment of transcortical inhibition* from some unknown source. His defect in respect of this abnormal motility can be nothing else, physiologically, than failure of one cortical element to inhibit another—a state of affairs the “extreme likelihood” of the occurrence of which is postulated by Sherrington for various fields of cortical action. The objective study here pursued precludes, as I have shown, the likelihood of the corticospinal system (Betz-cell to muscle) being voluntary in any exclusive sense, though it is the *usual* path by which voluntary movements are exteriorized. These must start at a still higher physiological level, as is apparent from considerations advanced in the previous section.

The involuntary movements of chorea are of a higher physiological type than those of Jacksonian epilepsy ; therefore even the transcortical part of the whole voluntary motor efferent system is on occasion usurped by involuntary action. "Volition," therefore—always speaking physiologically—is driven back to some other transcortical source, remote enough from the Betz-cell, whence its action on the whole motor path—say, fronto-rolandico-spinal—is, in the case of chorea, etc., very imperfect indeed ; or, it is driven out by involuntary cortical activity. Consideration of the phenomena of chorea and of athetosis, of tonic innervation and of apraxia, and of cortical reflex arc activity in general, in my view, clearly supports this line of argument. "*Volition, in short, seems to be little else than transcortical inhibition ; a "voluntary movement" little else than a movement that is "allowed" transcortically ; an "involuntary movement" (of this high class) little else than one that cannot be transcortically prevented.*"

It is of interest that the present studies point in the direction of these tentative conclusions, since different lines of research by other workers seem to indicate dissatisfaction with current conceptions. Lashley,⁵⁰ for example, says "the conception of volitional activity is too vague to have any scientific value," and holds that the "volitional" or "automatic" character of the centrifugal neural impulses of the cortex can be defined "only in terms of their complexity of organization and their relative importance in the total motor integration." Von Monakow,⁵¹ after declaring that the common views of pyramidal function must be essentially modified, says "it is probable that the pyramidal areas serve less for the execution of voluntary movements than for the inhibition of the kinetic functions of spinal coordination. Their function seems to be in the class of reflex activity." If lower centres are less under the directive than the inhibitory influence of higher centres, the same may well be true of some among the cortical centres themselves.

(3) Reverting finally, in this connexion, to the demonstrated disorder of reciprocal innervation characteristic of choreo-athetosis, we have to examine the question of the physiological level at which this phenomenon takes place. Reciprocal innervation is readily obtainable by stimulation of any particular single point on the motor cortex proper, but it is also produced by excitation of the internal capsule, the pyramids in the medulla, and at still lower levels. The action of decontraction of one muscle and excitation of its

opposite may certainly be elicited from the cortex, but, in Sherrington's ⁵² own words, "the seat of inhibition in these reactions from the motor cortex lies probably at the place of confluence of conducting channels in a common path, likely enough at their confluence upon the 'final common path'"—in other words, at the spinal level. Clinically, too, it is likely, from a consideration of data that I cannot now elaborate, that the link between the muscular components of a given movement (protagonists, synergists, and antagonists) develops its activity at the spinal level, but this is not the whole question. We know that reciprocal innervation is typical of numerous spinal reflexes elicited by stimulation of dorsal roots; this has been performed *seriatim* for cervical and lumbar roots by Sherrington and by Page May,⁵³ and the widespread movement patterns thus obtained are systematized and largely invariable. Without reciprocal innervation such spinal reflexes would be impossible. But from the cortex itself, as we have seen, similar co-ordinated movements can be obtained, although they are limited in extent, as has already been remarked. Whether precisely the same co-ordinating mechanism in the cord is used both for spinal reflexes and for cortical impulses is undetermined; according to Graham Brown,⁵⁴ "there is a finely differentiated mechanism in the motor cortex which does not set into action spinal centres in the same manner as do the in-going nerve-impulses carried by the sensory nerves of a limb, but is able in some way to pick out and set into action small portions, as it were, of the simple reflex movements." Be this as it may, *since reciprocal innervation is obtainable from the cortex, it can be deranged from the cortex.* It is in fact completely deranged in so-called tonic innervation, as I have previously demonstrated, and in these cases the lesions are situated in the frontal region and a transcortical effect is exercised on the pyramidal system. It is also disorganized in choreo-athetosis, as has here been fully exemplified, and the argument which I have developed points definitely in the direction of a cortical arc being utilized for the hypermotility of these clinical states.

As I have already indicated, the cerebellar element in afferent cerebello-cerebral defect of regulation is probably responsible for the disarray of component units in a cortical movement, "involuntary" as the latter is (in the cases under consideration), and some experimental support for this view is forthcoming in the recent researches of Tilney and Pike.¹¹ Their conclusion that the

cerebellum appears to play a prominent rôle in maintaining the proper relation in the "synergic units" of the body has not a little to be said in its favour.

Conclusions concerning Choreo-athetosis

My summary, in respect of choreo-athetosis, is that it represents a complex type of involuntary movement, for the carrying out of which motor mechanisms of cortical site are requisite. *No single and invariable anatomical site for lesions underlying its development is to be expected; it is the expression of disorder of a SYSTEM.* To its continuance afferent cerebello-cerebral defect of regulation contributes, and transcortical ("volitional") control over its manifestations is very imperfect. As I have on several previous occasions strongly urged, *any theory attributing its origin uniquely to striatal destructive lesions is impossible.*

These views of mine, here again developed in some detail and supported by a variety of arguments, at least in one respect have received recent confirmation. During the last year or two Minkowski⁵⁵ and Littmann⁵⁶ have adduced pathological and experimental evidence rejecting the striatal theory; they admit the complexity of the factors and the impossibility of attributing the phenomena to single lesions of single tracts at one and the same place invariably.

(For references, see list at end of Chapter XI.)

CHAPTER XI

DISORDERS OF MOTILITY AND OF MUSCLE TONE, WITH SPECIAL REFERENCE TO THE CORPUS STRIATUM

V. INVOLUNTARY MOVEMENTS AND THEIR PATHOGENESIS, *continued*. TREMOR. GENERAL SURVEY OF STRIATAL MOTOR FUNCTION.

Clinical Characters of Organic Tremor. Conditions influencing Tremor. Clinical Varieties. Nature and Pathogenesis of Tremor. The Problem of Voluntary Control over an Involuntarily Décontrolled Mechanism. Conclusions concerning Tremor. General Conclusions on the Motor Functions of the Corpus Striatum.

By comparison with chorea and athetosis, tremor is a motor derangement of a much simpler character. It may be defined as a succession of similar movements at equal intervals—more precisely, as a more or less regular, rhythmical, alternating contraction of a muscular group or groups and their antagonists. This definition however is not as precise as it appears, for the clinician encounters very fine and quick tremors, felt rather than seen, in which the alternation is difficult to determine, while other rhythmical conditions are more elaborate than ordinary tremor, and others still closely resemble the latter superficially yet have a different origin. Various complex alternating movements of an involuntary sort have been grouped by the French under the term of “rhythmias,” but this of course is more descriptive than definitive. For the purposes of this Chapter, attention will be confined to tremors whose organic origin is matter of general acceptance.

Clinical Characters of Organic Tremor

(1) *Rate*.—Doubtless some tremors are slower than others, yet the rate of the periodic muscular contraction is fairly constant, averaging from four or five to seven or eight a second. This rate

is not apparently a function of the length of the muscles concerned, for careful measurements in cases of paralysis agitans will as a rule give the same figure for short muscles such as the interossei or other small hand muscles and for larger ones such as those of the neck or trunk. Tremors of wide excursion are not of necessity slower; if anything, they are sometimes rather faster than tremors of narrow limits. From a study of numerous examples of striatal disease I calculate the average rate at six a second, as near as may be. This rate is found to vary within moderate limits at the instance of factors of both an extrinsic and an intrinsic kind.

(2) *Range*.—Every intervening grade from the finest range to a coarse and wide movement may be met with; and in the same patient the excursion often varies from time to time, sometimes

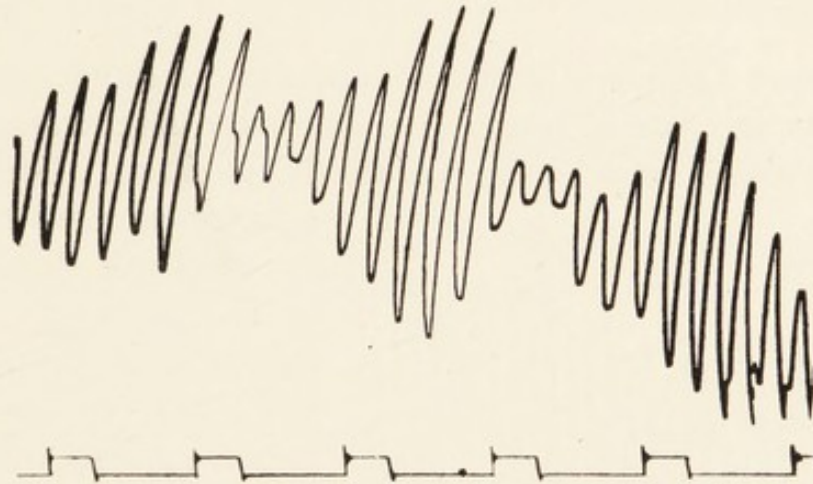


FIG. 34.—Tremor in Parkinsonism. Variation of amplitude with a certain regularity.

in a regular, sometimes in a highly irregular way. Fig. 34, taken from a case of postencephalitic Parkinsonism (unilateral), exhibits this range variation clearly, and a fair degree of regularity in the variation. Irregularity of range excursion is seen in Fig. 35 (also from a postencephalitic case).

(3) *Rhythm*.—On the whole, the majority of organic tremors exhibit a rhythm from which they depart but rarely; the tremor is set in such and such a way, and modification seldom occurs. The rhythm of the hand movement in the Parkinsonian from whom the tracing of Fig. 36 was secured is almost mechanical in its perfection. Fig. 37 represents another Parkinsonian tremor, of equally clock-like rhythm but of variable amplitude.

(4) *Localization*.—While theoretically any set of skeletal muscles and its opposite group may be seized upon by tremor, the small

muscles are more particularly its seat. A mere catalogue of tremor localizations would serve no useful purpose; what is of greater interest is the curious way in which tremor will shift its incidence

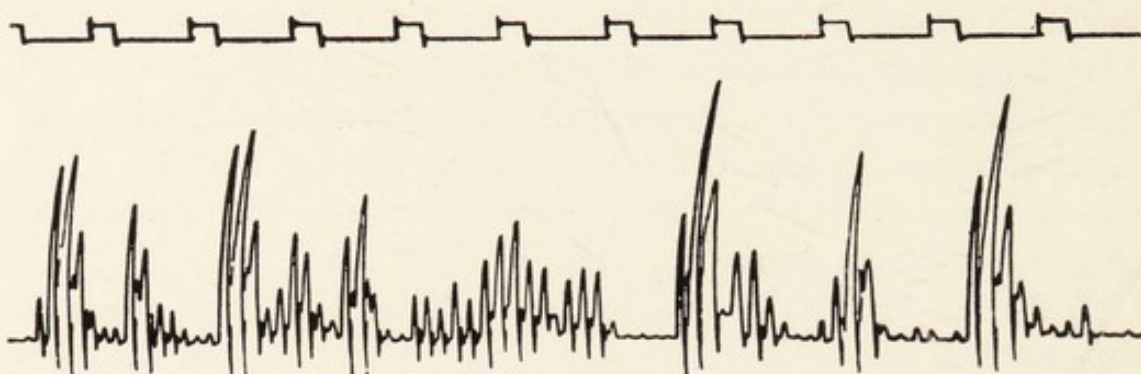


FIG. 35.—Parkinsonian tremor. Highly irregular amplitude.

in a moment from one group to another. I have noticed this particularly in cases of progressive lenticular degeneration, but it is a feature also of not a few cases of paralysis agitans. For example,

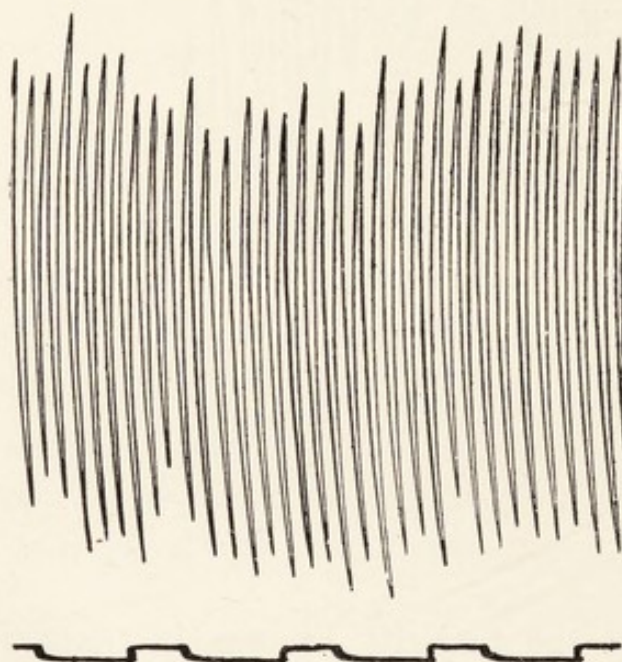


FIG. 36.—Parkinsonian tremor: almost mechanical rhythm.

a flexor-extensor tremor at the wrist will change to one of a side-to-side character (radial-ulnar deviation), or give way to one of pronation-supination, and so forth. Again, it sometimes will migrate in a still more extended fashion, leaving the arm, or its distal segments, to pass to the head or trunk, or to the opposite side of the body. The records reproduced in Figs. 38 and 39 are from a case of progressive lenticular degeneration with a pronounced tendency of this kind for

the tremor to wander. Within a second or two of placing the tambours in position for a record of a wrist flexion-extension tremor it shifted its incidence to the pronator-supinator mechanism, but returned a few seconds later to the original group, with reduced amplitude (Fig. 38). On another occasion (Fig. 39),

a rhythmical and seemingly unvarying pronator-supinator tremor altered its character as soon as the tracing started, and eventually left the group altogether, passing to the biceps-triceps combination.

This variability or fluidity of distribution is suggestive not of a fixed structural disorder so much as of a morbid process prone to

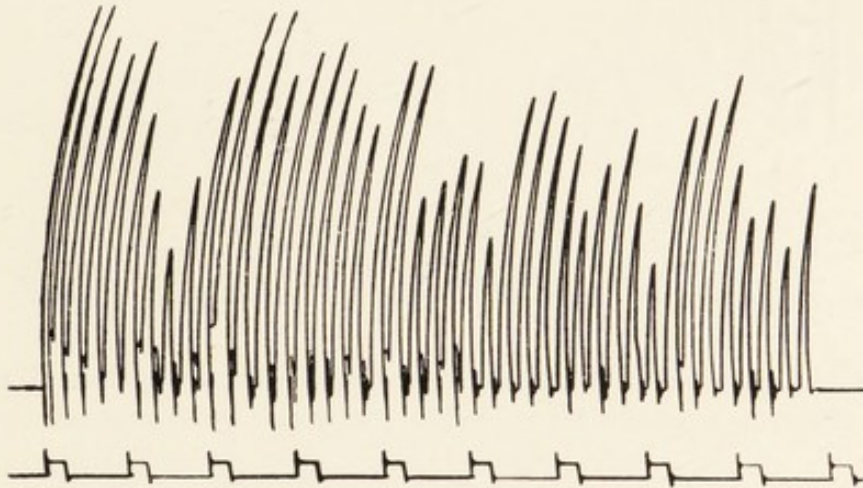


FIG. 37.—Parkinsonian tremor. Regular rhythm but variable amplitude.

develop itself in such and such a muscular combination as a sequel to the interjection of some additional factor, the nature of which will be discussed a little later.

A more remarkable illustration of this fluctuation is provided by a phenomenon which I have now observed some four times in

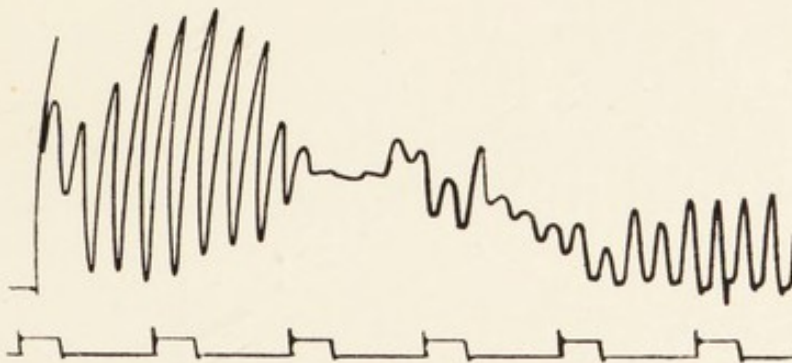


FIG. 38.—Progressive lenticular degeneration. Shifting of incidence of tremor.

cases of unilateral Parkinsonism, in each instance following epidemic encephalitis. The particulars of one case may be given.

It was that of a youth of 19, with left-sided Parkinsonism of a fairly severe character; his left hand and arm exhibited a typical rhythmical tremor which became particularly noticeable if the limb was unsupported, e.g., held out in front; support at all the joints either reduced it materially or caused it to disappear. On the right (normal) side was no sign of pyramidal or

extrapyramidal disturbance. The right arm could be held out by itself with absolute steadiness. Now, when *both* arms were outstretched the left at

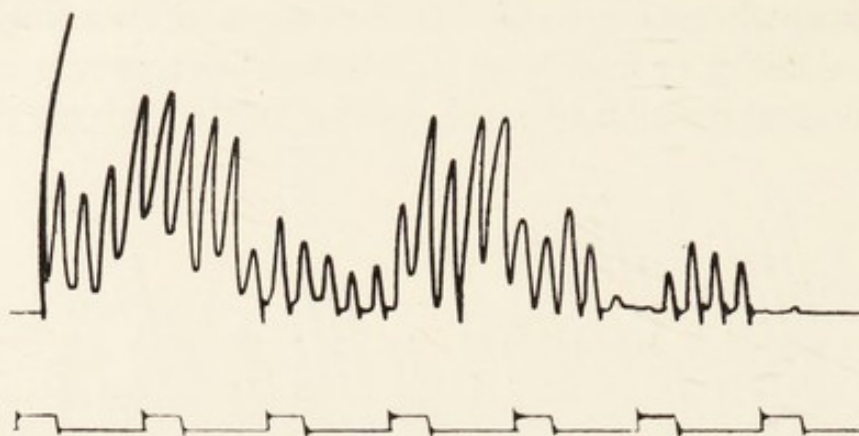


FIG. 39.—Same case as Fig. 38. Shifting of incidence of tremor.

once exhibited marked rhythmical tremor of flexion-extension at the wrist, and invariably, a second or two later, the right outstretched and unsup-

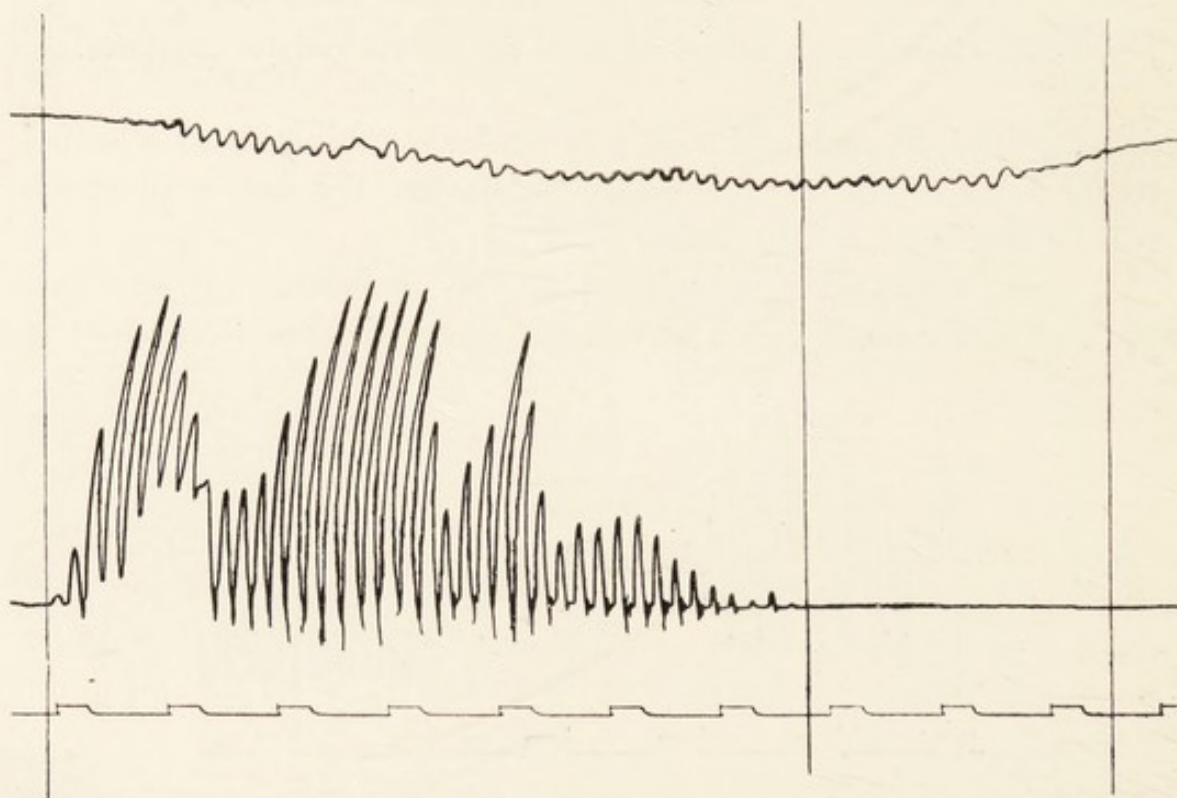


FIG. 40.—Unilateral postencephalitic Parkinsonian tremor. Temporary appearance of tremor on the *unaffected* side (above).

For details see text.

ported (normal) hand went into a similar but much finer tremor. Support applied to the left limb cut short its tremor, and within two or three seconds the fine right tremor would then disappear (Fig. 40).

Thus a limb in every objective respect apparently normal developed a tremor whenever its fellow of the opposite side, rather

severely involved in Parkinsonism, developed tremor; its tremor followed on the appearance of the Parkinsonian tremor, and ceased with cessation of the latter, yet both beginning and end always occurred a second or two later in the case of the former. Its range was much finer, but its rate and other characters were identical. This phenomenon is of considerable interest, furnishing, as it does, additional proof of the fluidity of many organic tremors—a feature to be taken into account in respect of pathogenesis.

Conditions influencing Tremor

We should derive a false notion of organic tremor if we contrasted it only with the known changeableness of “functional” tremors,

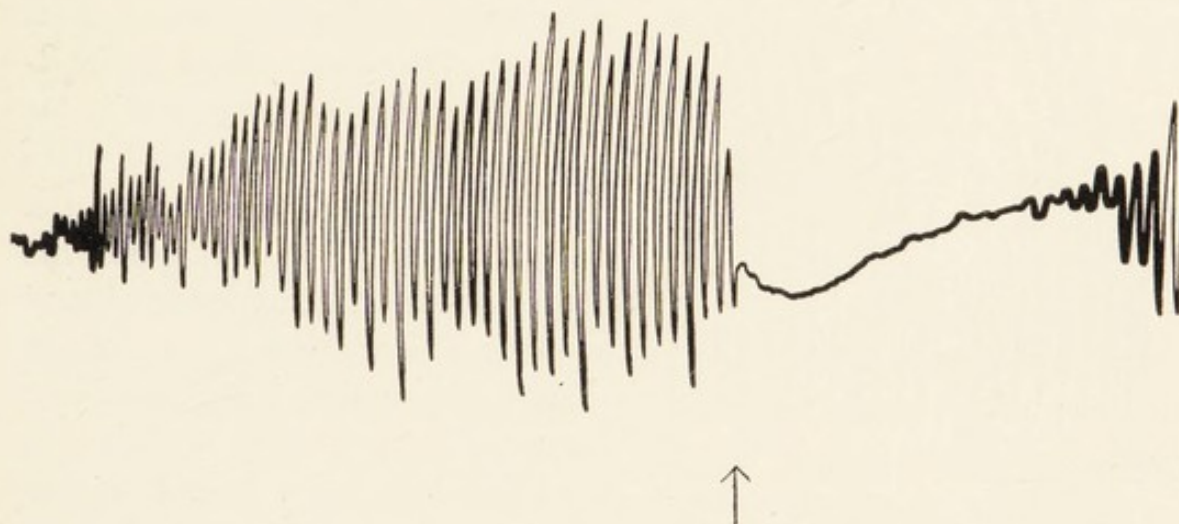


FIG. 41.—Parkinsonian tremor. Cessation after momentary cutaneous stimulus.

imagining change to be typical of the latter and fixity of the former. Numerous “functional” tremors vary in minimal degree, while organic tremors can be and are influenced by a whole series of factors, as will now be exemplified. When the back of the hand of a Parkinsonian patient whose tremor is in full swing is momentarily pricked with a needle the tremor may cease for many seconds. Fig. 41 is the tracing from such a case; at the arrow signal the dorsum of the moving hand was pricked for a moment and in this instance something like ten seconds elapsed ere it recommenced. (The crescendo character of this Parkinsonian tremor should be noted; the point is referred to below.)

Other forms of extrinsic excitation may produce an effect. At the arrow signal in Fig. 42 (idiopathic paralysis agitans) the sudden loud bursting of a paper bag filled with air caused no more than

the briefest disturbance of the rhythmical tremor, but sometimes the effect has been more pronounced. Stimuli of an emotional sort may or may not have their influence ; attention in this respect is a disturbing factor. I have always obtained better results in the maintenance of tremor for recording purposes when the subject's eyes have been closed. Not a few patients discover for themselves means of checking the involuntary movement, if only for a time. One of my patients could at once cause his tremor to cease completely by the simple procedure of supinating his affected forearm and hand. In another case, hyperpronation had precisely the same effect. This checking action did not escape the acute

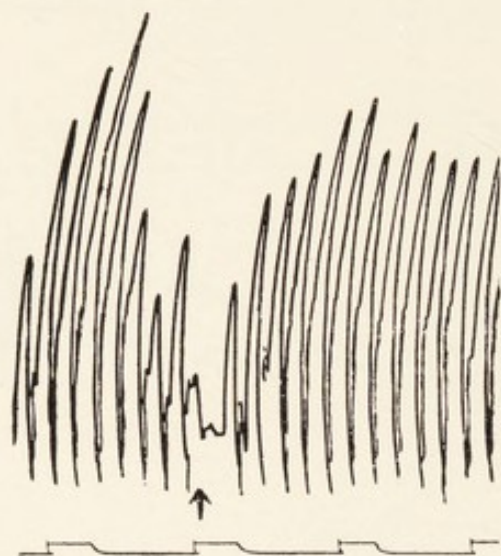


FIG. 42.—Parkinsonian tremor. Transient inhibitory effect of auditory stimulus.

observation of James Parkinson, who says in his Essay that "the wearisome agitation is borne until beyond sufferance, when by suddenly changing the posture it is for a time stopped in that limb." He cites the case of a Parkinsonian patient, who, "to illustrate his observation as to the power of suspending the motion by a sudden change of posture, being then just come in from a walk, with every limb shaking, threw himself rather violently into a chair, and said, 'Now I am as well as ever I was in my life.' The shaking completely

stopped ; but returned within two minutes' time." In other instances, however, it vanishes for reasons as elusive as they are inconstant.

Clinical Varieties

For years clinicians have recognized certain distinctions between the tremor of paralysis agitans and other striatal affections and that known as intention- or action-tremor,* the latter being characterized by the fact that it begins only with voluntary innervation and increases with continuing action or "intention" up to a point, whereas the former is not so increased but may, on the contrary,

* Recently a distinction has been drawn by de Jong⁵⁷ between action-tremor and intention-tremor : I have followed Ferrier in regarding the terms as synonymous.

diminish or cease. Further, action-tremor is seen only during voluntary motion, while the first variety occurs also when the limb is at rest. The supposed distinction has been recognized for centuries; it was already old when James Parkinson referred to it, quoting with evident approval the views of Sylvius de la Boe and Sauvages, and indeed the germ of the idea is discoverable in the pages of Galen. In Parkinson's words: "If the trembling limb be supported, and none of its muscles be called into action, the trembling will cease. In the real shaking palsy the reverse of this takes place; the agitation continues in full force whilst the limb is at rest and unemployed; and even is sometimes diminished by calling the muscles into employment."

Notwithstanding this authority, the distinctions are more apparent than real; some cases of disseminated sclerosis, for instance, definitely show what I may call a "rest-tremor" as well as an "action-tremor," while in progressive lenticular degeneration, as was demonstrated in my original monograph, the tremor is increased by voluntary action to a pronounced degree, though it is also prominent in the resting state. Thus combinations occur and invalidate distinctions. Moreover, in the finger-nose test the Parkinsonian limb at first often exhibits diminution or cessation of the tremor as it goes into action, yet after a moment or two of rest on the nose the fingers again become agitated, although the innervatory action of the limb musculature continues and the limb is certainly not "unemployed." Clinical observation proves further that while in some instances a Parkinsonian tremor is checked by firm voluntary closure of the fist of the affected limb, in other cases the effect is transient and minimal, and the tremor redevelops notwithstanding the active "employment." The exaggeration of the tremors of progressive lenticular degeneration by voluntary movement is strictly comparable to what obtains in disseminated sclerosis—an organic state to which "intention-tremor" is by some inaccurately, as I think, confined.

Reduction or cessation of tremor by supporting the limb—a reputed characteristic of action-, as compared with Parkinsonian, tremor—is met with nevertheless in the latter disease on occasion. Its occurrence has been shown in Fig. 40, taken from a typical case of the postencephalitic variety. On the other hand, the tracing of Fig. 43 is derived from a case of what I believe to be senile tremor. In this instance the tremor is seen solely during unsupported action of the limbs and vanishes instantaneously with support.

On occasion, Parkinsonian tremors exhibit a crescendo feature (cf. Fig. 41), which is reminiscent of the similar crescendo of action-tremors.

Finally, in this place, we may express doubt whether any hard-and-fast line can be drawn physiologically between a resting and an active state of muscle; action-currents in respect of these two differ only in a quantitative way. Clinically it seems a matter rather of degree of innervation than of differences in kind. From consideration of the data the conclusion is, that *the two types tend to run into each other and that neither is pathognomonic of involvement of a particular mechanism, radically distinct from that of the other.*

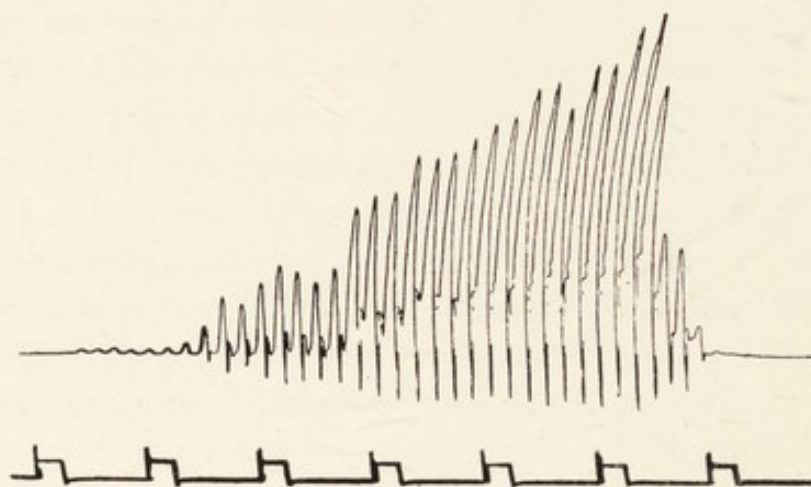


FIG. 43.—Tremor (organic) produced when limb is unsupported and ceasing when it is supported.

Nature and Pathogenesis of Tremor

From its nature we must allow that tremor can only be a phenomenon of a low physiological order, essentially different from the elaborate spontaneous movements of chorea and of athetosis. Tremor is purposeless and seems to have no "meaning," at least none easy of recognition, whereas choreic and athetotic action is subjectively purposeful if none the less futile.

(1) Various neuromuscular mechanisms are highly organized in the nervous system; others are organized in low degree or scarcely at all. Superior mechanisms are capable of great variation; inferior mechanisms are largely systematized. The more highly organized a mechanism, the lower is its physiological position. Spinal reflexes—e.g., the scratch-reflex—cannot be modified in respect of rate or character by variation of stimulus; cardiac and respiratory mechanisms are set at such and such a pace and, left to themselves, vary

within narrow limits only. The fifty-a-second rhythm of motor nerve impulses is practically a constant. By analogy, organic tremor is a physiological phenomenon of high organization and low variability, for as we have seen its fundamental characters alter little and its average rate is fairly constant. It is constituted by a rhythmic discharge of neural energy, appearing in pathological states—a release-phenomenon set in a particular rhythm the innate nature of which, and its *raison d'être*, seem at present to be quite uncertain.

Clonus, similarly, consists of a rhythmical alternation in contraction of muscular groups; is a physiological state of a low order; falls into approximately the same rhythm as tremor; and undoubtedly tends to manifest itself when disease sets free inferior mechanisms. That tremor and clonus rates of contraction should not vary greatly from each other cannot be a pure coincidence, even though the anatomical cause of their respective releases be structurally different in each.

(2) This neuromuscular rhythm being normally held in check by inhibition and demonstrable only in certain diseased or abnormal states, the next point is whether for its appearance a particular condition of low motor centres, or of the muscles themselves, or both, is requisite.

It is not found clinically in states of complete paralysis. A paralysed and flaccid limb is never the seat of tremor. On the contrary, relative integrity of motor power is essential, and if as the result of an additional lesion a limb exhibiting tremor is temporarily paralysed, one of the clinical concomitants is temporary disappearance of the involuntary movement. A fair number of cases are on record of the disappearance of tremor after an ictus of a hemiplegic kind, but it is important to note the absence of the tremor corresponds only to the time during which by diaschisis the *lower* motor neurones are functionally out of action; with some restitution of their proper activity, though the hemiplegia remains, the tremor will reappear. I⁵⁸ have myself reported a case of this kind, in which syphilitic mesencephalitis was associated with the clinical symptom of tremor; a stroke led to its cessation, but only for a time corresponding to complete outfall of function throughout the corticospinal system; the movement reappeared when this acute stage, implicating *lower* motor neurones by diaschisis, was over. One hundred years ago James Parkinson observed a case of paralysis agitans where a sudden stroke paralysed the right side

of the body ; " during the time of their having remained in this state neither the arm nor the leg of the paralytic side was in the least affected with the tremulous agitation ; but as their paralysed state was removed, the shaking returned." I am not aware of any recorded case where tremor has *permanently* disappeared after an ictus involving the pyramidal tract, and that alone.

(3) As a corollary, for the appearance of tremor clinically the muscles implicated must be in a certain state of tone. Atonic limbs do not exhibit the phenomenon ; they may be hypotonic, however, and, as a fact, also, either ordinarily tonic, or hypertonic. We must therefore ask ourselves whether any relation or proportion can be shown to exist between the manifestations of tremor and of muscle tone.

Of the common simultaneous occurrence of tremor and of muscle rigidity no question can arise ; paralysis agitans is a frequent and convincing instance of their coexistence ; on the other hand, we have to explain the coexistence of tremor and hypotonia in not a few cerebellar cases. To take the former first : My studies lead to the conclusion that, clinically speaking, *those cases of Parkinsonism that are most rigid show least tremor, or tremor that is most limited in amplitude.* I have been able to convince myself of this interrelation by the frequent observation, in postencephalitic cases, that with diminution of rigidity by treatment the patient will likely pass through a stage in which tremor becomes correspondingly more obvious. Several patients have made this observation independently and have commented on it. Conversely, tremor wide in excursion and sometimes almost violent is seen in cases where rigidity is at a minimum. Further, in any ordinary case of the syndrome those muscular groups which are more rigid (proximal segments) always exhibit less tremor than those that are less obviously rigid (peripheral segments), and the same remark is true in respect of lips, tongue, etc. Similar observations have been made for other striatal conditions such as progressive lenticular degeneration. Precisely those cases have exhibited the " wildest " tremors in which muscular rigidity has not been extreme.

Taking, next, cases where we have reason to suppose cerebello-mesencephalic lesions exist, rhythmical tremor resembling that of Parkinson's disease may be associated with a hypotonic state of the musculature. A number of such cases have been personally observed, and others have been made the subject of study by Holmes ²⁸ in his Croonian Lectures (1922). I am not able to cite

any clinical instance of such tremor having been modified by subsequent alteration of the hypotonia through additional disease to a hypertonia, though were such a case to be studied and reported it would prove valuable.

According to a view expressed by Holmes, tremor is seen in hypotonic states only when "both the agonists and their opponents are contracted," as when a finger is to be held steadily close to the nose without actually touching it. Diminution of postural tone allows the limb to oscillate, failure of fixation being followed by alternating contractions of certain groups of muscles and their antagonists. His explanation, however, only applies to cases where a hypotonic limb is being held voluntarily in such and such a position without support. Now we have seen similar oscillatory tremors to develop in limbs that are not hypotonic, as in some cases of paralysis agitans; and in progressive lenticular degeneration, where also the musculature is hypertonic, analogous tremors have often been seen. Again, some cerebellar tremors of a regular, rhythmical sort are seen when no active innervation is taking place, as when a limb is hanging over a chair or over the edge of the bed (Holmes). That voluntary innervation is not a factor in the spontaneous tremors of resting limbs is of course obvious; nor is gravity a consideration, for the limb may be so placed as to counteract its action, yet the tremor continues.

Evidently, as tremors that are clinically indistinguishable can be seen to develop in limbs whose musculature is of differing degrees of tone—hypertonic or hypotonic—no explanation based solely on the state of tone can be applied universally. It is commonly understood, I think, that the rhythmical alternating discharges of clonus are dependent on enhanced muscle tone in the units concerned; an abrupt passive stretching effects a stretching-contraction, and this in turn will bring about a similar contraction in the antagonistic group, and so on almost indefinitely, so long as adequate tension is passively maintained. No explanation of this kind is applicable to the spontaneous tremors encountered by the clinician. Nor is any elucidation based on the presence or absence of active innervation sufficient to cover all the facts.

While, then, as regards diseased states characterized by both tremor and rigidity the degree of the latter is usually in inverse proportion to the extent of the former, we are not justified in claiming the tremor to be an expression of the muscular state per se. It is never found in the extremes of absolute atonia or hypertonia, but

apart from these it may invade muscles whose tone varies within rather wide limits. Hughlings Jackson considered tremor and rigidity as it were opposite aspects of the same thing; he said that "tremor differs from rigidity, not fundamentally, but in degree," and explained the latter as being tremor "run together." He applied the idea in several other directions; for example, he spoke of a "diluted convulsion" as producing a condition "superficially like tremor." Again, he said that if by some magical process the movements made by a man in running were to occur simultaneously, "the result would be, not any movements properly so called, but an arrest of locomotion by a stiffening of the body into one attitude, a contention of all movements of locomotion." The applicability of this ingenious conception to the present question will be at once apparent to the reader. Walshe,⁵⁹ however, declares the view that tremor and rigidity are interrelated to be "wholly untenable," although he does not mention it as being due to Hughlings Jackson. That they are sometimes thus related is clear from their occurrence in inverse ratio, as illustrated above in Parkinsonian cases; but in other instances no such proportionate connexion is demonstrable.

(4) The fact that *tremor is not found clinically in either cortical or spinal cases* I consider of prime significance. A lesion of the corticospinal system is not by itself sufficient to produce tremor, as every one knows. As far as I am conversant with the literature, no case of tremor is on record pathologically in which the lesion has not involved grey matter situated somewhere in the vicinity of basal ganglia, mesencephalon, or cerebellum. In pointing out the occasional temporary cessation of established tremor by a corticospinal lesion, I do not admit the possible inference that, as Hunter states, "impulses through the cerebrospinal tracts are involved in the production of spontaneous tremor," if thereby the *cortical* arc is thought to be essential for its appearance. On the contrary, the evidence points in a quite other direction.

Brief consideration of the above-mentioned general localization of the lesions associated with the appearance of tremor clinically at once suggests the existence there of some prespinal (supraspinal) centre or centres of control over a lower mechanism, whose activity expresses itself in the form of rhythmical alternating contractions of a given muscle group and its opposite, at a more or less set rate. Following the invariable rule, we say that since the lesions associated with tremor are destructive the supraspinal centre controls or inhibits, and when it is injured the lower mechanism is "let go."

The alternations of tremor are of course an instance of reciprocal innervation, contractions of opposite muscles succeeding each other harmoniously in a never-ending series. We thus reach the conception that for tremor a particular condition of low motor centres is requisite, and that the state of tone of the muscles themselves is of less significance. None the less, it does not seem to be a mere coincidence that the levels of the lesions associated with tremor are also those from which alterations of muscle tone in large degree arise.

Magnus⁴⁷ has not seen tremor in any of his experimental thalamus animals, and comments on this absence of involuntary movement apropos of current striatal theories. Now in that animal *both* corticospinal paths *and* corpora striata are cut off from the parts further back (lower down)—an experimental state not in any way comparable to the lesions usually associated with development of tremor in man. In fact, few if any experiments hitherto have satisfactorily reproduced the kind of case the clinician frequently encounters. That tremor, none the less, is occasionally producible by experimental methods is known from the work of Economo and Karplus.⁶⁰ In Graham Brown's stimulation experiments on the mesencephalon (decerebrate animal) tremor has been obtained in the red nucleus vicinity. I had the opportunity of seeing one of the hemi-decerebrate cats of Bazett and Penfield,⁶¹ and observed tremor in the advancing of the limbs on the affected side. The now rather older experiments of Ferrier and Turner⁶² on the superior cerebellar peduncle have lost none of their importance *quâ* the experimental production of tremor.

(5) Clinicopathological evidence does not at present enable us to pronounce with certainty on the relative claims of corpus striatum and cerebellum (naming only two collections of grey matter) to constitute that tremor-controlling supraspinal centre which in view of the data here collected and the argument advanced we are justified in postulating. In former communications an attempt was made, possibly somewhat rigidly, to connect tremor with the lenticulo-rubro-spinal path. I have never doubted, and further pathological knowledge only serves to substantiate, the general conclusion formerly reached as to the localization of the lesions underlying its appearance; their relative limits have already been specified. But a purely clinical study of the present kind cannot give us all the material requisite for final decisions.

Viewed from another angle, however, as we have seen, tremor is

to be considered the outcome of activity of a low-grade physiological mechanism released from a *non-volitional* prespinal centre, and it therefore becomes a matter of some interest to ascertain the relation of volition to, and its degree of control over, this *otherwise* decontrolled (say) mesencephalospinal mechanism.

The Problem of Voluntary Control over an Involuntarily Decontrolled Mechanism

The question, it will be realized, is a particular instance of an interesting physiological problem of which other examples readily suggest themselves. When a given centre is under the inhibitory influence of say two other centres of different kinds, one volitional

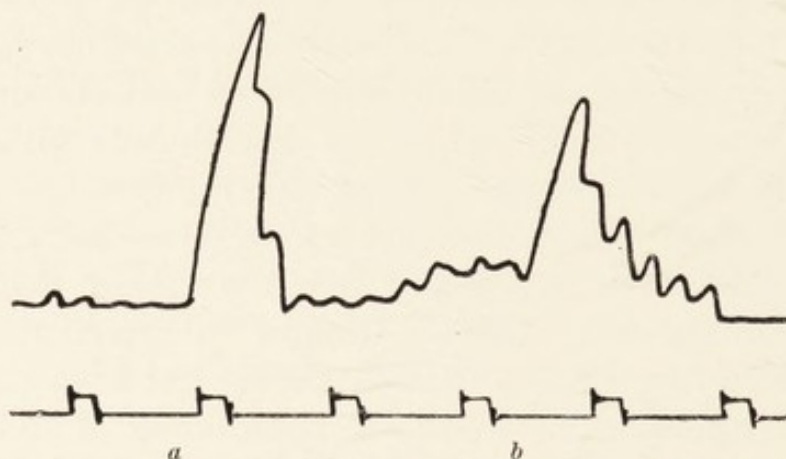


FIG. 44.—Parkinsonian tremor inhibited during “voluntary” contraction of affected muscles.

and the other non-volitional, what difference, if any, does the withdrawal of the latter control effect in the manifestations of the former control?

In the matter of tremor, voluntary influence is variable, fleeting, and not to be foretold. Thus during voluntary innervation of a tremor-exhibiting group of opposite muscles (say flexor-extensor) the current may suffice to blend the involuntary innervations into a tetanizing contraction, of one or other muscle, as may be, with corresponding (relative) disappearance of the phenomenon for the time. Yet this is by no means constant, for in other instances volition augments and exaggerates existing tremor, or, alternatively, seems to bring out a latent state of this, existing as it were in potentiâ. Attention is directed to the accompanying figures. Fig. 44 is a tracing from the left quadriceps extensor cruris of a Parkinsonian patient with moderate rigidity and well-marked typical tremor. At *a* and *b* voluntary contractions of the quadriceps

were made, and during the ascent of the curve (shortening of the muscle) no tremor contractions are visible, though these begin again at once with decontraction, and continue. Now in the case of another equally characteristic example of the affection (Fig. 45) no such checking of the involuntary contraction is seen; on the contrary, it is traceable both in ascent and descent of the curve (contraction and relaxation of *extensor communis digitorum*). If the reader will look back at Fig. 17 he will see how existing tremor is inhibited during voluntary contraction of finger extensors and begins at once when the summit of contraction is reached, while during synchronous and equally voluntary (in the sense that the whole movement is one of "volition") decontraction of wrist extensors the tremor maintains itself. The involuntary contractions blend with the voluntary contraction, but not with the voluntary relaxation that is going on simultaneously.

No other conclusion, I submit, can be drawn than that voluntary control over an involuntarily decontrolled mechanism is both imperfect and uncertain. It is unable to prevent the phenomena from making their appearance. The amount and maintenance of control are alike so variable that they preclude the possibility of similar fixed structural defects being responsible for the phenomena in every case. In a given instance it seems difficult to say what the interrelation of higher and lower mechanisms will turn out to be; tremor may be transiently inhibited by voluntary employment of muscles, or, on the contrary, it may accompany such action and refuse to be controlled—and this in different examples of the same disease. I believe considerations of this kind enable us to understand better the essential fluidity and variability of the clinical manifestations of tremor as described in preceding sections.

Conclusions concerning Tremor

Summarizing the results of the foregoing analysis of tremor, I may indicate some conclusions.

Tremor cannot be due solely to a particular or specific manifestation of muscle tone, for it is met with in hypertonic, tonic, and



FIG. 45.—Parkinsonian tremor not inhibited during "voluntary" contraction of affected muscles.

hypotonic states; the fact of its being set in a rhythmical way at an average rate of fair constancy suggests it is the expression of an inherent property of neurocellular activity; its mechanism is of low physiological grade and is controlled by an ("involuntary") prespinal centre or centres at the general level of corpus striatum, cerebellum, and mesencephalon.

Voluntary inhibition of this involuntarily decontrolled mechanism is essentially imperfect, inconstant, uncertain, and unsustained.

Tremor differs in fundamental characters from choreo-athetosis, above all in the fact that the law of reciprocal innervation is not abrogated in its case. Its underlying mechanism is distinct from that of the other group.

Tremor is an involuntary movement belonging to the old motor system (infracortical, subpallial), while choreo-athetosis is an involuntary movement of the new (cortical, pallial) motor system.

Motor Functions of the Corpus Striatum

No pretence has been made in these studies to cover the whole field of striatal symptomatology, since they have been oriented from a somewhat different viewpoint. Much of the research, however, has an obvious bearing on theories of the function of that particular collection of grey matter, so that some discussion of our analyses of motor and muscle tone disorders is desirable in this connexion.

At the outset the serious obstacles in the way of reaching final conclusions were enumerated. Our uncertainty as to the diseases to be designated striatal (and solely striatal) on precise neuropathological grounds was specially emphasized, and hesitation in respect of these is still both natural and warrantable, notoriously as far as Huntington's and Sydenham's chorea is concerned. By comparison Parkinson's disease and progressive lenticular degeneration may be supposed to be less equivocally striatal.

Clinical analysis of various types of movement, and of the precise symptomatology of the two last-named affections, gives the following results. The assertions that they are characterized by "loss" of associated movements, reaction and defence movements, and movements of co-operation, have been shown to be inaccurate. The view assigning a localization of "automatic" movements to the corpus striatum is equally devoid of foundation. The law of reciprocal innervation holds for movement in striatal cases of the above group. Catatonic phenomena have no place in the syndrome.

Still other points indicative of misinterpretation of alleged striatal motor symptoms have been recapitulated at various stages in the course of the studies.

It may be of interest, perhaps, to refer now to one or two fallacies that have been noted in regard to movements and their meaning and localization.

(1) One of the more serious is that which draws a hard-and-fast line between "voluntary" and "automatic" movements. No such line as a fact exists. "Automatic" movements, it will be granted, have been originally acquired, and, consequently, have at one stage contained a definite "voluntary" element. The child learns to walk. The supposition that this class of movement is at some unknown time transferred physiologically from a cortical to a striatal locus is devoid of any sort of probability and unsupported by any sort of evidence. However "automatic" walking and arm-swinging become, they remain faintly "voluntary." As I have previously urged, the gradation is not from "voluntary" to "automatic," but from "least automatic" to "most automatic," or from most to least "voluntary." After permanent lesions of the pyramidal system many acquired "automatic" movements are permanently lost. Were the intact corpus striatum the seat of these, they should exteriorize themselves via the lower motor neurone, which is not impaired per se in hemiplegia.

The other side of this question, so to speak, is that I have shown many cortical movements do not contain the element of "volition," but otherwise are similar in every respect to "voluntary" acts. Complex and elaborate tics are an instance in point, as are the motor phenomena of chorea. Before epidemic encephalitis became a clinical commonplace, tics were on their intrinsic clinical and etiological characters with entire correctness attributed to disorder of cortical function. Some writers now appear to envisage all motor symptoms through an encephalitic fog, arguing for a striatal localization of peculiarly elaborate movement-complexes because the virus of encephalitis has a striatal site in Parkinsonian cases! Such a claim can be made only at the expense of utterly ignoring the intrinsic clinical features of the movements in question. The corollary is logical, that if postencephalitic tics and bradykinetic syndromes are a sequel to striatal destruction, they must require for their activity *non-striatal* mechanisms. No hint is given as to where these are situated; but, assuming from the complexity of the movements that they are of cortical type, the inference would

be that the corpus striatum is higher in the physiological hierarchy than the cortex itself, and that it controls the latter, so that when it is disordered cortical activity is "let go." This is contrary to all our conceptions of cerebral physiology.

The argument in preceding pages has pointed in the direction of a transcortical failure of inhibition in the case of high-type movements such as those of chorea, and the same argument is applicable to tics and similar involved and intricate movement-combinations. Continuing this line of discussion, I have already alluded to the fact that movements obtained experimentally from the motor cortex proper are not identical in all respects with the movements of "volition." Physiologically, they are of a lower class. The clinical phenomena of Jacksonian epilepsy bear out this view. I have seen individual muscles in contraction from disease of the motor cortex, muscles that cannot be innervated "volitionally" in the same way. For example, I have seen in a Jacksonian case twitching of the right forefinger and thumb (a usual symptom, and of course not inherently different from "voluntary" movement of the same part) with *simultaneous twitching of the right levator menti alone*—this latter illustrates the present point. Again, I have observed distinct and prolonged *unilateral* twitching of the rectus abdominis in a Jacksonian case. I do not require to adduce more data to show that morbid processes can effect individual muscular contractions from the motor cortex, and that movements physiologically coupled can be dissociated similarly by *cortical* motor disorder. Further, interference with and derangement of the harmony of action of component muscular units in movement may occur at the cortical level and have been fully illustrated.

The Betz-cell anatomical area, then, is no more than a midway station between what I should prefer to call the true motor area of "volitional" action—viz. some much larger cortical region, possibly the frontal—and the lower centres of the pons, medulla, and cord. Clinical analysis of choreo-athetotic symptoms has shown us every variety of physiological dissociation and dysharmony in contraction of opposite groups, and converging lines of evidence have suggested such disintegration may take place at the cortical level.

(2) A second fallacy is common and insidious, consisting in the tendency to attribute to lesions at one physiological level symptoms which may in fact arise at more than one. If integration of neural

levels is essential to harmonious motor activity the possibility of disintegration through a series of levels must be steadily borne in mind. I have illustrated the point in the case of such symptoms as micrographia, akinesia, poverty of movement, and catatonia. Since such symptoms may be caused by dysfunction at more than one neural level, no exclusive apportioning of the particular symptom to an anatomical lesion on one only can be justified. For several Parkinsonian symptoms I have shown above the probability of the intervention of a "conscious" factor. Not a little of the seeming akinesia follows from the patient's unwillingness to initiate movements that he knows will entail an effort. "Awareness" has been proved to play a part in the meagreness of the reaction- and defence-movements of some Parkinsonians. In these latter types the implication of cortical arcs is highly probable, since the selected excitations reach the sensorium. If they are to be set down to striatal activities alone, and their absence to striatal disease, we are faced with the assumption that striatal function is a conscious function, for which it appears to me no evidence whatever has been adduced.

(3) A third source of confusion in a difficult subject arises from the variability of striatal symptoms at different times and under different circumstances, of which numerous clinical instances have been provided in these pages. To identify symptoms with anatomical lesions thus becomes an unusually delicate and precarious business. As long ago as 1912 I laid stress, in discussing the pathogenesis of tremor and other involuntary movements, on the possibility of dynamic modifications of function without recognizable anatomical lesions. "Loss" of movement is a very elastic term. We have had more than one illustration above of the habit of arguing that "loss" of a movement justifies the supposition that the normal ganglion is the centre for precisely those movements which are considered to be "lost." What, then, is the anatomical state of the organ when movements are present at one time and absent at another?

I should like, in conclusion, to approach the problems that have occupied our attention from still another point of view.

(1) It is a priori highly improbable, and contrary to the dictates of common sense, that motor phenomena so conspicuously diverse and of such varying complexity as myoclonus, tremor, athetosis, chorea, tics, bradykinesia, torsion-spasm, micrographia, and palil-

alia, should one and all be set down to disorder of striatal function. When we remember the histological simplicity and comparative structural homogeneity of the corpus striatum, in contrast with the greater dimensions, much more intricate cyto-architectonic complexity, and far wider connexions of the rolandic motor cortex, the idea of attributing all these disturbances to striatal disease, and of crowding corresponding "centres" into that ganglion, becomes nothing short of ludicrous. And we have seen that this Betz-cell area itself is physiologically inferior to a still larger motor region, which cogent evidence suggests may be constituted by the frontal lobes. Too little attention has been devoted by striatal enthusiasts to the phenomena of apraxia or dyspraxia, to the movements of co-ordinated epilepsy, to the syndrome of tonic innervation, to *epilepsia partialis continua*, and to other cortical and transcortical disturbances exteriorizing themselves in motor symptoms, some of which at any rate are of a kind persistently attributed to affections of the corpus striatum.

In this connexion an anatomical look-round will help to correct distorted perspectives. The corpus striatum is a much simpler morphological structure than the cerebellum, simpler even than the optic thalamus, in which nuclear organization has reached a stage unknown to its neighbouring ganglion. But no neurologist has hitherto assigned to disease of the cerebellum a heterogeneous miscellany of motor disorders of the kinds with which the corpus striatum is sorely overloaded. Some would reduce cerebellar function, perhaps with reason, to two or three elements, or even unify it completely. How, then, a relatively simple cell-fibre structure like the corpus striatum can mediate functions ranging from language (cf. *palilalia*) to innervation of muscle fibre (cf. *myoclonus*) is on the face of it inexplicable.

(2) Consideration of the intrinsic clinical features of some of the motor disorders put down to striatal dysfunction proves that under no circumstances can they be conceived of as resulting from defect or destruction of identical or similar mechanisms, for while some are physiologically of a high order others are of a low. Some can be understood only on the view that they contain elements belonging to the cortical series.

(3) My clinical studies have impressed me again with the belief, for which much support has been furnished here, that striatal symptomatology and striatal function must be comparatively simple; and they have forced me to the conclusion that the un-

wieldy and top-heavy symptom-complex which has gradually come to be erected on the ganglion must be rigorously stripped of non-essentials. In the preceding pages have been indicated the symptoms which must thus be discarded.

I have found no reason to modify in any important respect the syndrome originally enunciated, to the effect that the main features of disease of the corpus striatum consist of disorder of muscle-tone regulation and appearance of involuntary movements. These being "positive" symptoms, their causation is the freeing of non-striatal mechanisms from striatal control. The corpus striatum is a body one of whose functions is tone-control, for in its diseased state tone over-develops in the skeletal musculature generally; another of its functions is that of inhibiting the physiologically lower function of neuromuscular rhythm, for when it degenerates tremor is prone to make its appearance. I do not think, however, that in either respect its function is unique and exclusive of that of other centres of approximately the same order.

The "negative" symptoms of striatal disease would be constituted by actual loss or outfall of—what? Clinical analysis will not allow me to point to a single class of movement which is as a fact *persistently* and *completely* lost when the ganglion is out of action. We have had many instances of the ways in which different classes of movement are *hampered* as a sequel to striatal disease, but none of an absolute paralysis of any class. Fine movements of small muscles are seriously impaired—if the expression is preferred, "lost," but then they are "lost" also in pyramidal disease. In this, as in certain other respects, gaps in our knowledge of striatal function remain to be filled.

Throughout these studies no attempt has been made to assign separate functions to the composite parts of the whole ganglion, for in this region in particular speculation has outrun both clinical and pathological facts. On a previous occasion I have assembled the arguments against this special variety of striatal conjecture (see page 136).

These studies were undertaken in the hope of making a further constructive addition to our knowledge of the basal ganglia, and if at the same time my results should seem rather of a negative or destructive kind as far as some current views of striatal function are concerned, I can only say that revision was sorely needed.

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CHAPTER XII

PATHOLOGICAL LAUGHING AND CRYING *

Definition and Delimitation. Clinical Illustrations. The Emotional Factor in Pathological Laughing and Weeping. Bearing of the Phenomena on the James-Lange Hypothesis. The Mechanism of Emotional Expression. Some Theories of the Motor Phenomena. Criticism of Nothnagel's and Brissaud's Views. Outline of a Suggested Theory. Application to Diseased Conditions.

The problem presented by abnormal emotional expression, in the guise either of exaggerated or uncontrollable laughing or crying, or, conversely, of paralysis (at least in part) of the same mechanism, has not attracted much attention in recent years; nor has detailed criticism been offered, or advance made, on the position adopted by Nothnagel¹ forty years ago in reference to the latter, or by Brissaud² thirty years ago in respect of the former. From time to time doubts have been cast on the tenability of the hypotheses, yet little of a constructive nature has taken their place. The time seems opportune for a revision of the whole question, and for offering a somewhat different explanation of the syndromes. Again, as long ago as 1884 Professor William James³ admitted with characteristic candour that if the hypothesis of the emotions propounded by him "is ever to be definitively confirmed or disproved it seems as if it must be by asylum physicians and nervous specialists, for they alone have the data in their hands." As far as I am aware, however, no attempt has been made thus to prove or disprove it in the light of neurological knowledge derived from clinico-anatomical facts, so that a contribution from this viewpoint also may not be without interest.

Definition and Delimitation

At the outset the ground must be cleared by specifying exactly what is meant, for the purposes of this Chapter, by pathological

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laughing and crying. In the first place, reference is made solely to organic nervous cases in which, as a consequence of a recognizable cerebral lesion or lesions, attacks of involuntary, irresistible laughing or crying, or both, have come into the foreground of the clinical picture. While a not unnatural result of severe or chronic nervous disease may be a general depression of spirits, favouring tearfulness and irritability and other manifestation of an emotionally altered psyche, such conditions are not here considered. The emotional outbursts of the hysteric and the facile moods of the neurasthenic, likewise, are foreign to our subject. Patients who suffer from cerebral arteriosclerosis, in particular those in whose cases other indications point to impairment of function of the basal portions of the brain from an *état lacunaire* or other vascular change, often exhibit signs of an abnormal emotional state, and while this is without doubt the direct result of the disease, the usually diffuse nature of the latter precludes its being utilized for topographical purposes. Occasionally, however, the symptoms make their appearance in an arteriosclerotic case after an ictus, or series of these, and in such examples the localizing value of the syndrome may be in no way inferior to that of other cases with a more restricted and clean-cut pathological process for a basis. We are concerned, then, with the occurrence of exaggerated, forced, involuntary, uncontrollable laughing or weeping—the *Zwangslachen* and *Zwangswainen* of German writers, the *rire et pleurer spasmodiques* of the French.

In the second place, organic cases in which conservation of voluntary facial movement is combined with paresis or paralysis of the same musculature for the involuntary movements of laughing or smiling, though rare, have long been known to the neurologist. This defect in the mechanism of laughing is clearly but a part, and it may be only a small part, of the total somatic expression of that particular emotional state, nevertheless as such it is deserving of close attention. Its significance will be duly considered, and a fresh explanation offered of its pathological physiology.

Clinical Illustrations

I. Among organic affections apt to be associated with pathological laughing or crying may be enumerated double hemiplegia, pseudo-bulbar paralysis and disseminated sclerosis; its appearance after a single hemiplegia has also been observed, and, as remarked above, it is of moderate frequency in certain stages of basal degeneration from diffuse vascular processes. The exact nature of the morbid

affection is of less importance than its site; tumour growths, infective conditions, vascular degenerations, provided they are appropriately situated, may produce the symptoms indifferently.

By way of clinical illustration some personal cases may now be cited.

1. Double Hemiplegia.—One case of crying and one of laughing may be selected.

CASE 1.—A woman of 57 had suffered from left hemiplegia for one year, when a second stroke occurred involving the right side. Ever since the latter the daughter remarked that her mother had become, as she put it, "hysterical," laughing and crying at nothing.

On examination the patient was seen to have a distinctly vacant, apa-



FIG. 46.—Case 1. Pathological crying in a case of double hemiplegia.

thetic facial expression at rest. She was able to move the facial muscles voluntarily on both sides, though there was slight weakness of the left corner of the mouth. On the slightest stimulus, even when the observer simply came to her bedside, she at once assumed a lugubrious expression, her mouth opened widely, and a long, almost noiseless bout of weeping ensued, lasting for many seconds, even minutes, at a time (Fig. 46). During this spasmodic crying both sides of the face moved equally, and the eyes suffused with tears. Laughing attacks were extremely rare in comparison.

CASE 2.—A man, age 67, had two strokes in the same year (1916); the first was on the right, with moderate aphasic disorder; the second was on the left, and comparatively mild. Since the first attack, and to an increasing degree since the second, he had exhibited characteristic involuntary laughing. Whatever the emotional stimulus, and however slight, he at once began to laugh, and laugh loudly. Thus on reading the war

news he would at once smile, and the more serious and anxious the news, the more he laughed.

On examination there was some voluntary facial paresis on both sides, especially the left, some dysarthria, and some dysphagia, but during the laughing the facial movements were in no way restricted. A double extensor response was present.

2. Pseudobulbar Palsy.—The syndrome occurs in this affection with greater frequency than in any other (Fig. 47). Hartmann⁴ reported eight cases, in all of which moderate or slight voluntary weakness of facial movement was associated with pronounced spasmodic laughing or crying, or both. Similar clinical examples have been recorded by Schaffer,⁵ Weisenburg,⁶ and many others. Instead of quoting personal cases I shall refer in some detail to an instance of the affection reported by my former teacher, the late Dr. Charles Beevor,⁷ which presents very unusual features. It so happens I am in a position to add some facts to Beevor's striking study of the case.



FIG. 47.—Pathological laughing in a case of syphilitic pseudobulbar palsy.

CASE 3.—A young man of 23, known to have been infected with syphilis, had three hemiplegic attacks, two involving the left side and one the right, as a result of which there was *complete* loss of all *voluntary* facial movements as follows: closing the eyes, elevation and retraction of the angles of the mouth, opening and closing the mouth; there was likewise *absolute* loss of *voluntary* biting, deglutition, phonation with elevation of the palate, and of voluntary inspiration and expiration (such as coughing). Both sides were affected equally. Contrasting with this profound degree of volitional palsy was preservation of emotional movements of laughing and crying. The patient was continually laughing; in fact, on the slightest provocation, or on none, he went off into rounds of laughter, which made examination of his facial and respiratory condition very difficult. Reflex movements of coughing, sneezing, and yawning were readily obtained, but in laughing the angle of the mouth was not retracted so well on the left side as on the right, whereas in yawning the facial movement was symmetrical. As the patient was unable to open his mouth, the house physician used to sit at the foot

of the bed and yawn deliberately. Eventually the former caught the infection and yawned automatically, whereupon the Sister of the ward promptly took the chance of popping food into his mouth.

A further illustration of the phenomenal *rire spasmodique* may be added to the account. About this time a peripatetic quack was touring the minor music-halls of London and claiming to cure all and sundry complaints by means of electricity. The patient went, and was shown on to the stage. Diagnosed as hysterical by the electric "expert," he was submitted to a series of violent and painful electrical applications, but the more they hurt the more he laughed, till at length he was quickly hustled off, and on his return the following evening was refused admission.

In view of its almost unique nature and significance from the standpoint of the mechanism of laughter and its localization, further reference is made to this case below.

3. Single Hemiplegia.—Many cases have been recorded of a single ictus precipitating the tendency to the symptoms under consideration (Brissaud,² Broadbent,⁸ Burzio,⁹ Mills,⁵⁸ and others). In some instances, doubtless, there is more widespread vascular disease than the single stroke would indicate, and in some tumour cases, similarly, there is more general disturbance of cerebral function. Yet the onset of the syndrome may undoubtedly coincide with the appearance of limb symptoms which are strictly unilateral.

CASE 4.—A woman of 50 had an attack of right hemiplegia and aphasia, and thereafter became peculiarly lachrymose, bout after bout of weeping succeeding each other all through the day, with copious tears. Thus my coming to her bedside was the unvarying signal for a fresh outburst of uncontrollable crying; in fact, whenever she was spoken to the same occurrence was noted. On examination there was slight volitional weakness of the right side of the face, but the asymmetry was not observed during the bouts of weeping. Before the hemiplegia no such emotional overaction had been noticed.

CASE 5.—A male patient, age 59, presented in a fairly typical form the thalamic syndrome in association with a left hemiplegia. In addition to corticospinal symptoms (extensor response, etc.), objective diminution to painful and thermal stimuli was found over the same side, with characteristic over-reaction; an athetoid attitude of the hand was also noted, and occasional involuntary movements of the same. He complained of constant and severe "burning pins and needles" in the left arm and left side of the face.

In addition, the patient presented the symptom of involuntary laughter in a marked degree. This had made its appearance after the stroke, but was greatly augmented when, some two years later, weakness of the right side, associated with some sensory change, began to develop.

Though involuntary laughing or crying is not regarded as a usual

or even occasional accompaniment of the thalamic syndrome, there is no reason why the two should not sometimes be combined, and the above is an instance in point.

By way of contrast, allusion may be made to a series of cases published by Féré,¹⁰ under the title of *Le fou rire prodromique*. In these the onset of irresistible emotional overaction preceded the development of hemiplegia. In the first case an elderly gentleman of 64 began to suffer from *rire spasmodique*, being convulsed with laughter over trifles. A few months later right, and subsequently left hemiplegia developed, death ensuing from pseudobulbar paralysis. The second case was that of a man, of the same age, in whom uncontrollable explosions of laughter from minimal stimulation were followed by an irresistible desire to sleep. These phenomena were of daily occurrence for some four months, when a severe left hemiplegia supervened. In view of the significant fact that thereafter all involuntary explosive laughter ceased up to the date of the patient's death, eighteen months later, the absence of exact neurological details is regrettable.

No case of this description has come under personal notice, but I have seen cases of disseminated sclerosis in which involuntary laughing was the first symptom to attract attention.

4. It is unnecessary to furnish clinical examples from cases of other organic affections of the nervous system, but an exception may be made in respect of *disseminated sclerosis*, because of the frequency of the syndrome in that disease.

CASE 6.—In the case of a man of 32, with typical symptoms, attention was first directed to the emotional change by the fact that when reading of a perfect stranger's death he would begin to weep; with the narration of amusing incidents exaggerated laughing would set in. Under observation bursts of long, uncontrollable, but almost noiseless laughter took place at the veriest trifles. In the course of my examination I asked the routine question whether he had any difficulty with the bladder, and replying in the affirmative, he added he had already "ruined four pairs of trousers," and went off into an apparently interminable series of peculiar hollow laughs, which convulsed the whole ward as well as himself. So facile became the mechanism that he would laugh whenever he began to speak, as though the stimuli of contracting muscles were sufficient to set it off.

II. We turn now to cases characterized by *volitional normality* and *emotional abnormality* of facial movement.

The occurrence of facial paresis or paralysis during emotional expression is a very old observation. Almost a century ago Sir

Charles Bell ¹¹ wrote : " As you find the portio dura in possession of distinct properties, all of them related to respiration, breathing, speech, and expression, you will not be surprised that these functions should occasionally be differently affected ; as, for example, a man will continue to possess the power over the nerve as the nerve of speech, and yet he will be incapable of expressing the usual signs in laughter or in crying. In short, you find that your patient sometimes exhibits paralysis of the side of the face only when he smiles or laughs, at other times it is not observable." An equally old and impressive clinical illustration is furnished by the case recorded by Stromeyer ¹² in 1837, concerning a girl of 12 years, " in whom the right side of the face continued expressionless in emotions, and showed no increased action in accelerated respiration after running, going up stairs, etc. Nevertheless, the child was as able to control the muscles on this side as those on the left ; she could move the angle of the mouth, dilate her nostrils, wrinkle her forehead, and contract her eyebrows at will. . . . On compressing the epigastric region, it appeared that the right half of the thorax scarcely took any part in the forcible (involuntary) thoracic respiration which was induced by the pushing back of the diaphragm. When this kind of examination was made, the apathy of the one half of the face was particularly manifested at the nares ; whilst the right one remained immovable, the left one expanded fully at every act of inspiration." This case is particularly informative and will be referred to again.

Numerous examples of the combination of volitional activity with emotional paralysis have since been recorded (Nothnagel,¹³ Bayerthal,¹⁴ Mills,¹⁵ Borst,¹⁶ Feiling,¹⁷ Monrad-Krohn,¹⁸ and many more). Of various examples that have come under personal observation three only will be cited.

CASE 7.—A young woman of 27 had suffered for six months from increasing headache, giddiness and attacks of vomiting. Her cerebation became slower and her memory poor. For about the same time weakness and paræsthesiæ of the left limbs had been noted.

On examination the optic discs were clear but hyperæmic. Objective evidence of a slight left hemiparesis (arm and leg) was obtained. When the patient showed her teeth, closed her eyes, etc., no paresis of the left face was discoverable, but on emotional stimulation a notable asymmetry was at once shown, the left side exhibiting a considerable degree of " mimic paralysis " (Figs. 48 and 49). The later course of the case indicated more definitely the presence of a cerebral tumour in the region of the right internal capsule and right regio subthalamica.

CASE 8.—A male patient of 41, known to have had syphilis, suffered from a stroke involving right arm and leg, and not long thereafter developed a right posthemiplegic hemitremor. On examination the left pupil was inactive to light and reacted poorly with convergence, whereas the reactions of the right pupil were normal. Lateral conjugate deviation was good in both directions, but upward and downward movement was very poor. Voluntary facial movement was normal on the two sides, whereas on smiling the right facial movement was minimal, and on laughing the difference was notably accentuated. That is, the right side of the face exhibited expressional paralysis.



FIG. 48.—Case 7. Normal volitional facial movements.



FIG. 49.—Case 7. Mimic palsy of left side of face in laughing.

CASE 9.—A girl, age 17, developed characteristic symptoms of intracranial tumour in the shape of headache, giddiness, vomiting and papilloedema. The presence of a double Argyll Robertson pupil, nystagmus, tremor, incoordination in finger-nose test (left), etc., pointed to a mesencephalic localization.

On volitional movement of the face no defect was observable, but on expressional movement (laughing) the left side of the face moved only slightly, whereas the right side moved normally (Figs. 50 and 51). At autopsy a tumour was found situated in the tegmentum and upper pons, involving the left side more than the right (Fig. 52). Both crura were normal.

It is worth noticing that in all the reported instances, as far as I have been able to ascertain, this paresis or paralysis of expressional facial movement has been on one side only. No bilateral case

seems to have been observed; none has come under my notice, and a similar remark has been made by Spiller.¹⁹

The Emotional Factor in Pathological Laughing and Weeping

The natural question that must arise for discussion is whether the emotional outbursts of which descriptions have been given reflect the mental state of the individual concerned at the moment of their expression. Are such overwhelming laughter and tears



FIG. 50.—Case 9. Normal volitional facial movements.

activated by appropriate stimuli, or do they in their turn induce the appropriate frame of mind, or have they any emotional content at all? To enable us to come to some decision the following considerations appear pertinent.

1. The stimuli are often inadequate and inappropriate. Instances have already been supplied of the truth of this statement. One patient (Case 4) cried when she was spoken to, when anyone sat beside her, when a hand was laid on her arm. In a case reported by Giannuli,²⁰ the patient, a man of 66, used to walk about the hospital with his eyes glued to the ground; if he so much

as raised them to meet anyone else's gaze he was immediately overcome by compulsory laughter, which sometimes lasted for four or five minutes. Brissaud² recounts the history of a patient of his, an intelligent hemiplegic, who was told incidentally by a lady that her little dog was dead; in a moment the fountains of emotion were opened; a mournful visage was succeeded by tears, tears by sobs, and sobs by a Rabelaisian effect on his sphincters. Another of Brissaud's patients, a student with syphilitic hemiplegia, was forced to abandon novel reading; "*les malheurs de l'héroïne le font*

éclater en sanglots, ses joies lui donnent de véritable transports." Reference has been made (Case 6) to spasmodic laughter accompanying the mere attempt to speak; Giannuli's patient, similarly, laughed as soon as he opened his mouth to describe the pain he suffered from renal disease. I have already mentioned how one of my patients (Case 2) laughed at grave war news.

In not a few cases, therefore, the emotional exhibition develops at the bidding of stimuli so minimal as to escape detection; in others, it is motivated by impulses appropriate enough if trifling, but it is excessive out of all proportion to the impressions originating it; in still others, a stimulus of a particular quality is followed by an emotional outburst of a contradictory sort, to which it is not appropriate. Intelligent patients suffering in this fashion are often conscious of the insufficiency of the psychological impulse, and the more intelligent they are the more painfully aware do they become of the incongruity in their affliction.



FIG. 51.—Case 9. Emotional or mimic palsy of left side of face in laughing.

2. From what has been said it will be understood that the apparent, visible emotion need not correspond to the patient's real feelings at the time—an observation often made. Oppenheim⁴⁷ says that "the patient has to laugh against his will, although his mood is not gay; this distresses him greatly." A patient of 63, whose case is given by Dupré and Devaux,²¹ a pseudobulbar with excessive attacks of laughing and crying, indicated by gestures of impatience and denial how much he was annoyed and ashamed at his performances, and how he suffered in mind at the constant caricaturing of his real feelings by their outward expression. A

patient of my own (Case 5) told me how one day his daughter had hinted plainly enough that she thought "Dad was putting it on a bit"; incensed at her unbelief, he rose from his chair to give her a box on the ears, but, his legs giving way, he had to throw his arms round her neck to keep himself from a fall, and in this (for his angry state of mind) ignominious position he burst into explosive laughter.

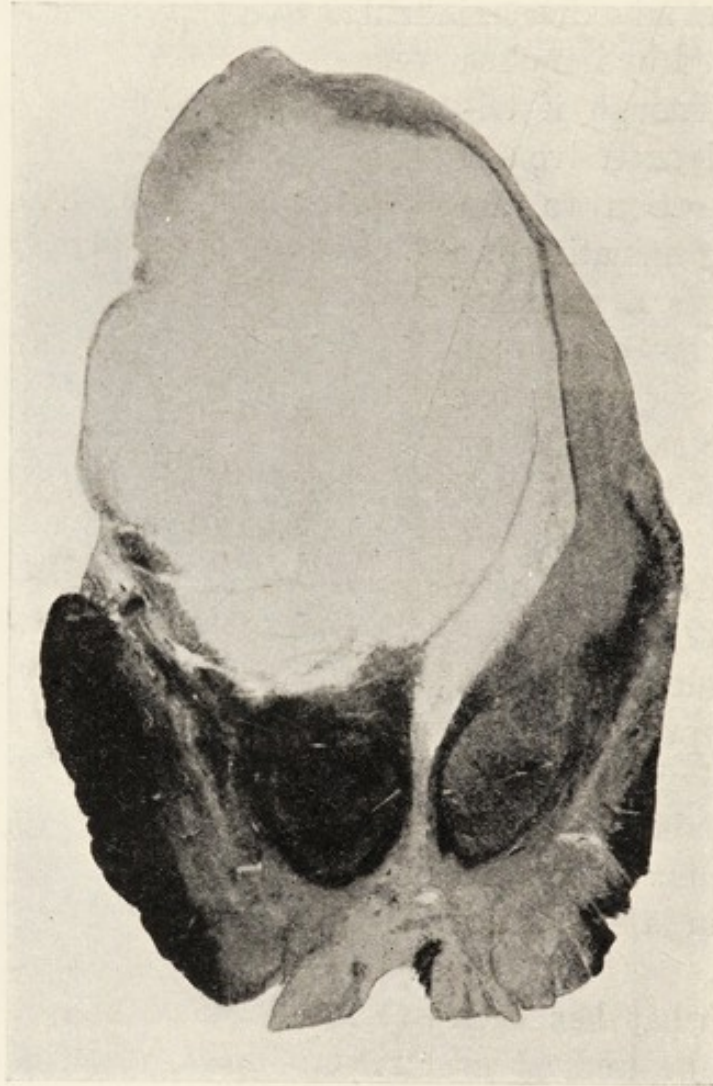


FIG. 52.—Case 9. Tumour of mesencephalon, involving mainly the left side, and interfering with the non-volitional faciorespiratory path through the tegmentum, on that side. Both crura, with their pyramidal tracts, were intact.

I have endeavoured to ascertain from intelligent patients whether when thus overcome by laughter against their will and in opposition to their real feeling they do not, in spite of the latter, end by experiencing the emotional state commonly associated with laughter, and I am satisfied it is not so, in some instances at least. On the other hand, Moutier ²² was informed by a young pseudobulbar patient that a single brief attack of *pleurer spasmodique* left him quite indif-

ferent, whereas prolonged and repeated bouts had the effect of saddening him and of bringing on tears legitimately motivated by the thought of the affliction under which he laboured.

Thus if the exaggerated laughter or weeping of the hemiplegic or pseudobulbar may, and often enough does, correspond to his emotional mood of the moment, it is not so always; we can scarcely suppose that inextinguishable laughter represented the feelings of the patient (Case 3) undergoing painful treatment at the hands of a showman.

3. As a further point, it is important to note in some instances the invariability of the emotional response, whatever the stimulus. Some of the sufferers can only laugh, others can only weep. Why this should be so is not easy to determine. Crile's²³ theory in respect of normal emotional activity is that both laughter and crying have the purposeful effect of utilizing released kinetic energy, and of "working it off" until it is neutralized. Hence one may pass into the other almost indifferently. In some pathological cases, none the less, the mechanism seems to be "set" for one only. On the whole, a rough generalization suggests that in cases of disseminated sclerosis the manifestation is one of cheerfulness; in pseudobulbars laughter and tears occur indifferently; in arteriosclerotic cases tearfulness seems to predominate. The difference in type, however, must not be taken too absolutely. Brissaud²⁴ gives an amusing description of a patient with extremely marked *pleurer spasmodique*, whose facies during the attack became "affreusement grimaçant." In the bed next to him was a patient with *rire spasmodique*, who used to roar with laughter at the weeping of the former, and this on occasion led the first to change his *pleurer* to an equally phenomenal *rire*, though the tears and the lachrymose physiognomy to some extent remained through it all.

At the same time, the emotional display is a genuine manifestation of feeling, which no one witnessing these attacks can doubt. Prolonged exhibition of every manifestation of grief, in facial expression, respiratory accompaniment, and secretion of tears, or, alternatively, equally patent demonstration of hilarity, in features, respiratory movement, rosy and suffused countenance, and tears too, it may be, is too definite to be mistaken for the mere shell of a mental state devoid of emotional tone. I cannot agree with Bianchi⁶⁰ when he declares that the "weeping and laughter of such sufferers are only simulacra of the real emotions." The display gathers impetus as it proceeds, and if it is initiated by trifling

stimulation, and in a comparatively cold emotional atmosphere, its avalanche nature is conclusive proof of the involvement of the entire mechanism, somatic and visceral, of emotional expression. In a word, it differs from normal emotional reactions solely in its inevitability, its frequency, its uncontrollable character, the occasionally contradictory relation of cause and effect, and the extreme facility with which it is induced ; in expression and accompaniments it is identical.

Bearing of the Phenomena on the James-Lange Hypothesis

The theory of the emotions associated with the names of Carl Lange and William James is too well known to require elaboration in this place. Since, however, it seems to some extent to be misunderstood, or, at least, incorrectly applied, I have taken the opportunity of re-reading the originals in a convenient reprint.²⁵

According to Lange, emotion is the product of (1) a cause—a sensory impression which usually is modified by memory or a previous associated image, and (2) an effect—viz., vasomotor changes, which in their turn produce changes in bodily and mental functions. He asks, “What lies between these two factors, or does anything lie between them?” His answer is that nothing lies between ; the bodily phenomena are aroused immediately by the cause, so that the emotion consists exclusively of these functional disturbances. “Take away the bodily symptoms from a frightened individual ; let his pulse beat calmly, his look be firm, his colour normal, his movements quick and sure, his speech strong, his thoughts clear, and what remains of his fear?” Whether a mental or a physical impression induces the reaction, the chief requisite for formation of an emotional state remains the same for both, viz., stimulation of vasomotor centres. “We owe all the emotional side of our mental life, our joys and sorrows, our happy and unhappy hours, to our vasomotor system. If the impressions which fall upon our senses did not possess the power of stimulating it, we would wander through life unsympathetic and passionless, all impressions of the outer world would only enrich our experience, increase our knowledge, but would arouse neither joy nor anger, would give us neither care nor fear.”

The views advanced so plausibly by James are not entirely identical. He does not postulate the intervention of stimulation of vasomotor centres, presumably those in the medulla, but rather holds that “particular perceptions produce widespread bodily

effects by a sort of immediate physical influence, antecedent to the arousal of an emotion or emotional idea." One of his arguments, as is known, is the speculative one of inability to picture an emotion without consciousness of all the feelings of its characteristic bodily symptoms. "What kind of an emotion of fear would be left if the feelings neither of quickened heart-beats nor of shallow breathing, neither of trembling lips nor of weakened limbs, neither of goose-flesh nor of visceral stirrings, were present, it is quite impossible to think. Can one fancy the state of rage and picture no ebullition of it in the chest, no flushing of the face, no dilatation of the nostrils, no clenching of the teeth, no impulse to vigorous action, but in their stead limp muscles, calm breathing, and a placid face?"

A third variant of the theory is that elaborated by Sergi,²⁶ who considers Lange's views too restricted, and includes in the mechanism other medullary centres for organic life than the vasomotor, viz., respiratory and vegetative or sympathetic centres.

The hypothesis is, in a way, described with slight inaccuracy as the *peripheral theory* of the emotions. Both Lange and Sergi assume intervention of bulbar centres *before* peripheral elements are set in motion; James, too, admits cortical activity *before* the periphery is reached by reflex currents. Yet all seem to be convinced that no emotion is felt in consciousness until the brain is in its turn again reached, this time by visceral impressions. James gives the following résumé of his position: "An object falls on a sense-organ and is apperceived by the appropriate cortical centre; or else the latter, excited in some other way, gives rise to an idea of the same object. Quick as a flash, the reflex currents pass *down* [*italics mine*] through their preordained channels, alter the condition of muscle, skin, and viscus; and these alterations, apperceived like the original object, in as many specific portions of the cortex, combine with it in consciousness and transform it from an object-simply-apprehended to an object-emotionally-felt."

Thus the first stimulus is ectoperipheral, followed by an endoperipheral stimulus, and emotions are not felt till the impression aroused by the latter reaches the cortex. But between these is an *efferent* impulse to viscera and certain skeletal muscles, so that an in reality somewhat complicated degree of neural activity, both central and peripheral, must precede awakening of the emotional feeling. A diagram from Kirchhoff²⁷ will render the idea more clear (Fig. 53).

Objections to the theory outlined above have been raised from different sides.

(1) Psychological difficulties have been emphasized by not a few who are competent to criticize. Sully,²⁸ for example, is convinced that the presence of an element of feeling at the very beginning of an emotional experience can sometimes be clearly observed. Pleasurable emotion can be started by "agreeable sensations," via eye and ear, and by the "agreeable perceptions" which grow immediately out of these. When we laugh at some absurd incongruity in speech or manners "the perception which starts the laugh is an emotional perception," and "is flooded from the very first with

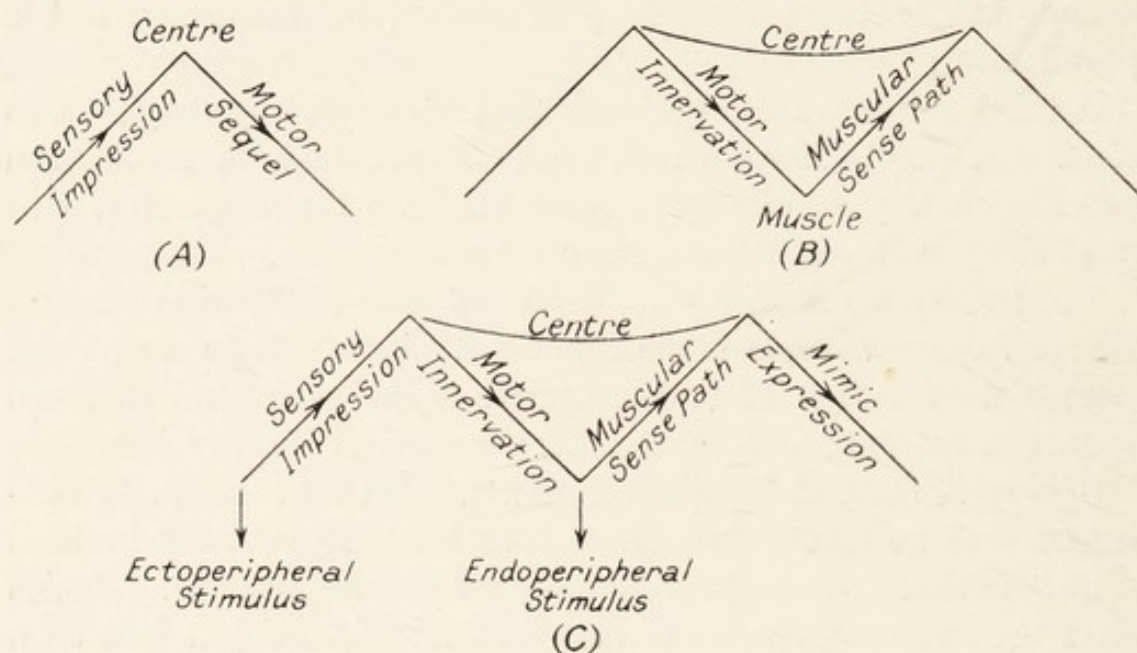


FIG. 53.—(Kirchhoff.)

A. Ordinary reflex arc. B. The so-called reflex circle of Bell. C. The reflex chain for the expression of emotion.

the gladness of mirth." Further, once we are exhausted with laughing at a comedian we may be physically incapable of any further manifestation of emotional feeling, and yet we may still feel the full appeal of his funny stories, of his amusing antics. The objection has been raised by Störring²⁹ that Lange and those who agree with him reduce emotions and feelings to sensations, even though they be complex sensations of a particular kind. He maintains that emotions represent fusions of organic sensations and affective elements, and that it is impossible to peel away the mass of organic sensations from the total in such a way as to justify the statement that nothing is left. To enter at large into these and other psychological criticisms is, however, outside my present purpose.

(2) Physiological objections have been formulated by Sherrington,³⁰ Cannon,³¹ and Bianchi,⁶⁰ among others.

Sherrington by appropriate experimentation removed completely the sensibility of viscera and of skin and muscles behind the shoulder in a number of dogs, yet this procedure resulted in no obvious diminution of an emotional character. "A mere remnant of all the non-projecting or affective senses was left, and yet emotion persisted." His conclusion is that organic and vascular reaction, though not the actual excitant of emotion, strengthens it. Cannon's researches have been devoted more particularly to an analysis of the visceral components of emotional states, and he has shown (in terror, rage, and intense elation, for instance) that the responses in the viscera "seem too uniform to offer a satisfactory means of distinguishing states which, in man at least, are very different in subjective quality." Since *various* strong emotions are expressed in the diffused activities of a *single* division of the autonomic system, the bodily conditions which have been assumed by some psychologists to distinguish emotions from one another "must be sought for elsewhere than in the viscera."

(3) The clinicopathological argument is the one to which attention is here specially directed.

Consideration of the clinical examples cited above shows, as I believe conclusively, that outward expression by no means always corresponds to the patient's real feelings. Though the emotional states are pathological, they are in quality identical with normal emotions. Yet more than one patient has protested against the laughter or tears being taken as the index to his actual affective state. The conclusion is unmistakable, that the bodily reverberation, as James calls it, is not *per se* the emotion; the latter is not, so to speak, the mental expression of the former. With all the outward appearances of mirth and hilariousness, and with concomitant activity of visceral mechanisms, the individual may not only not feel happy, but his state of mind may be in patent conflict with the apparent emotion. It is clear, therefore, that the James-Lange hypothesis must be materially modified if it is to be brought into line with observations such as have here been recorded, with no complete fusion between peripheral and cerebral components. The emotional framework may be activated without its afferent impulses being synthetized with the pre-existing mental state into a harmonious whole. A touch on the trigger releases the neural pattern into exaggerated action; yet the patient may remain

mentally detached, largely, if not perhaps always entirely, uninfluenced by somatic and visceral currents streaming centrewards. From the standpoint of the clinician, therefore, I find myself in accord with the physiologist when he declares that "the reverberation from the trunk, limbs, and viscera counts for relatively little . . . as compared with the cerebral reverberation to which is adjunct the psychical component of the emotional reaction." Indeed, some of my own and of the reported cases of others indicate the possibility of dissociation between psychical and physiological elements in the emotion.

Under normal conditions, practically all writers agree on the reinforcing and intensifying of emotional cerebral states by the advent of somatic and visceral impulses (cf. Mott ³²), but our study of certain diseased conditions of organic origin must lead us to accept with caution deductions as to the genesis of emotions made by the introspective method of the pure psychologist.

The Mechanism of Emotional Expression

In the expression of the emotions of joy and of sorrow, the only two with which we are here concerned, somatic and visceral factors are to be distinguished, though the latter, perhaps, are less in evidence or less intense in their activity than in other emotions that might be named. In the case of laughter there is, on the somatic side, involvement of facial and respiratory musculatures. It is unnecessary to describe the exact features of the former, familiar as it is to every one. As for the latter, automatic rhythm of the respiratory centre in the medulla is rudely interrupted by prolonged inspirations, followed by short and broken expirations. Coupled with respiratory movements are laughter sounds of laryngeal origin and of varying character and pitch. If laughter is overwhelming, other muscles beyond those of face and respiratory apparatus will be implicated; in fact, there may be a good deal of diffused movement, even to the extent of rolling on the floor. For our purpose, however, attention need only be directed to face and chest. On the visceral side, capillaries and arterioles are dilated; the eyes sparkle and increased glandular secretion is observed; the skin reddens and glows.

In striking contrast, as far as skeletal musculature is concerned, is the expression of sorrow; the facial movements of the latter are the reverse of those of laughter, while in respect of respiration there are short and interrupted inspiratory movements, succeeded by

prolonged expirations—again the reverse of the other emotion. Further, there is a general inhibition, face and chest apart, of the rest of the voluntary musculature. On the visceral or vegetative side a degree of hypofunction results from a widespread vaso-constrictor effect, according to Lange, and is explanatory of the pale colour, sunken features, sensations of cold, lassitude, etc., that accompany sorrow.

Now objective study of facial and respiratory movements is a *sine qua non* for understanding difficult problems connected with the question of mimetic centres as opposed to centres for voluntary movement, and of localization of lesions producing the clinical phenomena under discussion. The no less important matter of central representation of the visceral system is not at present, unfortunately, capable of the same objective examination.

A theoretical question not without practical bearing may be briefly touched on in passing. Which contributes more to the total emotional feeling, the facial and respiratory movement, or the visceral activity? Or are their respective quotas approximately equal?

The evidence I wish to adduce is based on investigation of pathological cases in which free movement of facial musculature is impeded by organic disease. It has been a routine matter in examination of such cases to inquire into the patient's feeling under the influence of appropriate stimuli. Among the material examined have been cases of facial diplegia, facial myopathy, myasthenia gravis, and of paralysis agitans and postencephalitic Parkinson's disease. The conclusion is that the patient can readily feel and be acutely conscious of experiencing a particular emotional state such as that associated with hilarity and joy in spite of minimal expression in the face. Moreover, the facial element may, as in the case of the "snarling smile" of myasthenia, be a positive distortion of normal movement, yet the feeling is in no degree lessened or altered. A facial diplegic, as one has often seen, may preserve a mask-like countenance and yet be moved by "inward" laughter. Romberg³³ mentions complete absence of expressional movement in one of his cases of facial diplegia, and says the patient "was very sensitive on this point, and termed it his greatest misfortune that he was forced to be joyful or sad without making any demonstration of his feelings to his fellow-creatures." Similarly, Sir Charles Bell quotes a case from Dupuytren's clinique, that of a girl of 16, with facial diplegia, whose countenance bore a serious char-

acter, contrasting forcibly with her frame of mind ; “ she retained her good humour and sometimes laughed heartily . . . as if behind a mask, her face being quite immovable and grave, whilst the emotion and sound of laughter prevailed.”

From cases of this kind it may legitimately be argued that the time-worn controversy as to the actor's feeling the emotional quality of his part by assuming a suitable facial expression can be dismissed in a few words. An artificial assumption of an emotional facies is practically a negligible element ; only when psychical and appropriate visceral components are fused can emotion be felt acutely, and the latter is less significant than the former ; mere portrayal of an emotion may deceive the audience, but never the actor himself. A little acquaintance with the observed facts of clinical neurology serves to emphasize the inaccuracy of the idea which Edgar Allan Poe ³⁴ puts in the mouth of one of his characters : “ When I wish to find out how wise, or how stupid, or how good, or how wicked is anyone, or what are his thoughts at the moment, I fashion the expression of my face as accurately as possible in accordance with the expression of his, and then wait to see what thoughts or sentiments arise in my mind or heart, as if to match or correspond with the expression.”

Some Theories of the Motor Phenomena

Clinical study reveals the existence of three types of inter-related motor disorder in connexion with emotional facial movement.

1. In the ordinary case of organic hemiplegia, the face on one side is paresed or paralysed for voluntary movement, but not for emotional expression ; there is volitional asymmetry, but involuntary symmetry. Careful examination sometimes shows that three, rather than two, stages can be distinguished : thus (*a*) we may observe volitional asymmetry ; (*b*) at the commencement of emotional movement, as in smiling, there may be involuntary asymmetry, the sound side moving before the other ; (*c*) when, however, laughing is well established the stage of involuntary symmetry is reached.

2. In the ordinary case of double hemiplegia or of pseudobulbar palsy we meet with a condition of affairs as in (1) above, except that the voluntary paresis or paralysis is bilateral.

3. The third group is constituted by the class of case referred to at the outset, where voluntary control is perfect, while emotional

facial expression is unilaterally paresed or paralysed; the condition is one of volitional symmetry and involuntary asymmetry.

An explanation sometimes given for the phenomena of the first group is that the facial musculature of the two sides acts normally in emotional expression as a physiological couple, and is presumably represented bilaterally in the cerebral hemispheres; hence unilateral lesions underlying hemiplegia will not throw out one-half of the pair. This view, known as Broadbent's hypothesis, is supposed to give an apt explanation of the relative conservation of voluntary movement in the upper face, as opposed to the lower, in hemiplegia, and has been widely applied in other directions, mainly in reference to *volitional* action. Its original formula ³⁵ is as follows: "Where the muscles of the corresponding parts on opposite sides of the body constantly act in concert, and act independently, either not at all, or with difficulty, the nerve-nuclei of these muscles are so connected by commissural fibres as to be pro tanto a single nucleus. This combined nucleus will have a set of fibres from each corpus striatum [read, cerebral cortex] and will usually be called into action by both, but it will be capable of being excited by either singly, more or less completely according as the commissural connection between the two halves is more or less perfect."

The application of this theory to the matter of conservation of mimic expression (a *non-volitional* movement) in unilateral facial hemiplegia may be criticized on the ground that it is going beyond Broadbent's original contention, but the criticism is not valid.

Consideration of the other groups, however, has naturally led to the idea of separate and distinct paths for emotional and for volitional facial movement.

1. As long ago as 1865 Saunders ³⁶ pointed out that the facial muscles have three distinct modes of action: as respiratory muscles, reflex; as muscles of expression, emotional; and as voluntary muscles in the strict sense. He postulated in the peripheral trunk of the seventh a distinct set of fibres for each of these different kinds of action, each connected independently with different excitor centres, so that one might cease to function through disease, but not the others. Long before him, Sir Charles Bell, as already remarked, had noted the different types of activity, and had offered an explanation that is not, perhaps, very clear. Bell ³⁷ says in one place: "We must determine whether even the portio dura of the seventh nerve may not lose one faculty and retain another. I suspect that the influence of passion, as those of smiling or laughing,

is lost in consequence of affections that do not destroy the entire power of the nerve." Elsewhere he declares that: "We really have no reason to conclude that the one property of a nerve requires a finer organization than another. I should rather suppose that this power of expression is constituted with a finer relation to the condition of the mind and of the body; and, therefore, we may suppose is more easily affected by slighter derangements."

Bell's view is probably not incorrectly described as a theory ascribing differences in function to *differences in degree of affection* of the seventh, and in this respect is classifiable, with that of Saunders, as a peripheral theory. The hypothesis is certainly untenable. If the trunk of the facial nerve is peripherally involved, it is involved for all modes of activity without any doubt. The only exception, or apparent exception, that I know of is furnished by Spiller,³⁸ who observed in some instances of pressure on the facial nerve from extracerebellar tumours that emotional expression may be more impaired on the affected side than volitional expression. This he considers indicative of "a certain stage of peripheral facial palsy," but he admits that "the impairment of facial emotional movement may result, in part at least, from the pressure of the tumour upon the medulla oblongata and pons." A converse condition seems to have been observed by Monrad-Krohn,³⁹ who says that "in some few cases of peripheral facial paresis one may once in a while find a faint suggestion of this dissociation, inasmuch as the emotional innervation seems to result in a slightly *stronger* [*italics mine*] movement than the voluntary innervation."

Since my acquaintance with Spiller's article, frequent examination of analogous cases has failed to reveal any instance corroborating the observation. It must be remarked, moreover, that as regards both Spiller's and Monrad-Krohn's cases the difference is one of *degree* only between voluntary and involuntary innervation, not one of preservation and loss respectively, or vice versa. The problem, it may therefore be taken, bears rather on the possibility of separate, central, paths for the two main varieties of facial action; in Sherrington's terminology, separate "private paths" converging on a "final common path."

NOTHNAGEL'S THEORY

2. In 1879 Nothnagel,¹ influenced by the much older conceptions of Sir Charles Bell and of Romberg, assumed that the simplest way to explain the ordinary motor phenomena in single hemiplegia,

where the face is unilaterally paralysed for voluntary but not for mimic movement, was to postulate the existence of a "psycho-reflex" facial path, distinct from the facial division of the pyramidal tract; he thought the optic thalamus and its connexions with the cortex were situated on this psychoreflex path, which in hemiplegia was unaffected. To explain the converse syndrome, viz., unilateral emotional paralysis with retention of voluntary control, he tentatively suggested that "perhaps in such cases there is a local lesion in the optic thalamus." This view, commonly referred to as Nothnagel's theory, has since been applied far and wide, but its pathological basis, by which alone it can finally be proved or disproved, has never been satisfactory. We must note that Nothnagel himself did not go so far as to place mimetic centres in the optic thalamus—the form in which the theory is usually expressed; his exact words, quoted above, indicate the uncertainty in his own mind as to the facts.

Cases of his own on which he subsequently relied ⁴⁰ are anything but unequivocal, as can be discovered readily if the originals are consulted. His views, however, have found support from Strümpell, Bruns, Bechterew,⁴¹ and many more, and have been repeated in successions of textbooks. Cases of thalamic disease thought to corroborate them have been published by Borst,¹⁶ Kirchhoff,⁴² Raimann,⁴³ and others, yet not many of these will stand rigorous investigation. In nearly all multiplicity of lesions renders them unsatisfactory and untrustworthy.

Hopeless as it is to disentangle specific mechanisms from widespread disorders of function entailed by equally widespread lesions, the *general* localization of these cases ought not to be ignored; a good example of involuntary crying caused by a thalamic tumour has been recorded by Weisenburg and Guilfoyle.⁴⁴ On the other hand, one or two records may now be cited in which the thalamus has been (presumably) intact. In the Dupré-Devaux ²¹ case of pronounced *rire et pleurer spasmodiques* a marked *état lacunaire* of each putamen was associated with softenings in the anterior limb of the left internal capsule, while the thalami were unaffected. Burzio's ⁹ patient, a woman of 24, suffered from severe left hemiplegia and from irresistible attacks of laughing; the lesion was a vast softening of the right lenticular nucleus, with involvement of the posterior limb of the capsule, while all its anterior limb fibres were degenerated; cortical softenings were also seen in right frontal and postcentral gyri. The thalamus was apparently normal.

Similar *rire spasmodique* in a case of disseminated sclerosis was attributed by Touche ⁴⁵ to the presence of plaques in the anterior and posterior segments of both internal capsules, in lenticular nuclei, etc., but no change was detected in thalamus or regio subthalamica. A considerable number of thalamic lesions are reported in the literature which have not been associated with symptoms such as are under discussion, and which need not here be particularized.

Lacking in precision, as must be the conclusions drawn from the data thus briefly sketched, the possible rôle of the thalamus, i.e., of some part of it, in the production of involuntary laughing and crying is not to be lightly dismissed, as we shall shortly see.

BRISSAUD'S THEORY

3. Another hypothesis was advanced by Brissaud.² According to him, *integrity* of the thalamus is essential for the appearance of spasmodic laughter or weeping; the causative lesion is one involving the anterior limb of the internal capsule, in that part where he places his *faisceau psychique*, or frontothalamic tract of control over thalamic centres. One of his cases was that of a man of 48, with right hemiplegia and complete aphasia; extremely marked compulsory weeping was an additional feature. Post-mortem, softening was found in the left putamen, extending across the anterior segment of the internal capsule; symmetrical lesions were discovered in the right putamen, but these only touched the anterior limb of the capsule. Many small cortical lesions were also noted, though their exact position is not given. In the case recorded by Giannuli ²⁰ and used by him to support Brissaud's hypothesis, there was a considerable softening in one anterior capsular limb, yet both thalami also were patently the seat of degenerative disease—in contradiction of Brissaud's contention, just mentioned, for integrity of the thalamus in spasmodic laughing and crying.

In his lectures on pseudobulbar paralysis Brissaud ⁴⁶ appears to place centres for involuntary expression of emotion in the basal ganglia ("noyaux opto-striés"), and to argue that the syndrome we are concerned with is a product of irritation: "il signifie toujours une irritation capsulaire" (anterior segment). He regards the phenomena as on a par with the tendency to spasm shown in cases of hemiplegia: "si les centres en question ne sont pas détruits mais simplement excités par une lésion de voisinage, ils traduisent le spasme hémiplégique en déchaînant le rire et le pleurer."

Notwithstanding its plausibility the hypothesis will not bear serious investigation. Apart from primary objections to the view that the symptom is irritative and not of the nature of a release-phenomenon, Brissaud relies for support of his thesis on anatomical connexions of cortex with corpus striatum and on anatomical views of the ansa lenticularis which have not been confirmed by subsequent investigation.

Oppenheim,⁴⁷ who with Siemerling described exaggerated laughing and crying in pseudobulbar palsy in 1886, supposes it due to "lesions of the centres or interruption of the tracts which have an inhibitory effect upon the bulbar centres," without specifying further the exact position of one or the other. He states, moreover, that these facial movements may be affected in every possible way "according as the morbid foci have an irritating or a paralysing effect." Here again, we think, the student is offered a theory which is a little nebulous.

4. Reference has been made above to the work of Hartmann⁴ on pseudobulbar paralysis. Arguing from the experiments of Bickel, who observed the explosive character of voluntary movements in animals after the production of sensory ataxia by appropriate cortical lesions, Hartmann is inclined to regard the absence of centripetal impulses as a factor in pathogenesis. He believes that a defect on the *afferent* side through the optic thalamus is responsible for the release of involuntary emotional activity. In this respect he is in agreement with von Monakow,⁴⁸ and, in a way, with Lewandowsky⁴⁹; the latter argues that peripheral sensibility has a greater influence on mimic expression than on voluntary facial innervation, and that since the optic thalamus is only a "Schaltstation" of sensibility on the way to the cortex, and not an autonomous organ, lesions productive of amimia or of expressional overaction may be situated there, though they may not; they may also occur in the cortex or in subcortical fibre-systems. He is doubtful whether mimic reactions can be obtained at still lower levels. The opinion expressed by Bechterew⁵⁰ is that the syndrome is dependent on two factors: (1) removal of voluntary control, and (2) the influence of exaggerated involuntary stimuli. Elsewhere he states, negatively, that *rire spasmodique* is certainly not due to implication of the voluntary paths to the facial muscles.

From these and from other writings that might be cited but that do not call for detailed reference, the student of the subject will appreciate the truth of Lewandowsky's final comment, that more

exact observations are required, and that the hypotheses usually advanced are characterized by indefiniteness and absence of precision.

Outline of a Suggested Theory

In order to aid understanding of a possible theory explaining both amimia and mimic overaction, it is desirable to indicate the way in which, as I conceive it, the subject should be approached.

1. Omitting in this place consideration of the visceral components of laughing and weeping, I think it imperative to note the participation of both *facial* and *respiratory* mechanisms in the act of laughter (or weeping), both physiological and pathological.

The physiological association of facial and respiratory musculatures in the expression of emotion scarcely calls for any comment, so obvious is it. Bell called the seventh the "facial nerve of respiration"; paralysis of the lower face (mouth and nose) was described by him as "paralysis of the respiratory functions of the facial." Implication of the face in sneezing, facial spasms occurring with respiratory gasps in extremis, collaboration of the facial apparatus with the other in ordinary breathing and speaking, are simple instances of the action of this important synkinesis. For simplicity's sake, we may allude to it as the faciorespiratory mechanism. We note that its normal activities are involuntary, i.e., it is under voluntary control only to a limited extent. Laughter may be stifled, tears may be restrained, no doubt; practice may enable the individual to inhibit its function to a varying degree; ordinarily speaking, however, the faciorespiratory mechanism works "on its own," whether the circumstances be physiological or pathological.

The localization of the "nœud" of this mechanism is still uncertain; we have to postulate a centre linking the seventh nucleus in the pons with the motor nucleus of the tenth (nucleus ambiguus) in the medulla and the phrenic nuclei in the upper cervical cord, etc. By all analogies this centre must be supranuclear; for the sake of argument we may suppose it has an upper pontine site.

2. Our second preliminary consideration is to bear in mind the existence and function of the respiratory centres proper, for ordinary automatic breathing, situated in the medulla. With their normal action must also be associated co-operation on the part of larynx and face, otherwise normal breathing might partake of the noisy character observed in various diseased conditions.

The most recent work on localization of respiratory centres is that of Lumsden,⁵¹ who has shown, by numerous experiments on cats, rabbits, dogs, and monkeys, the somewhat elaborate nature of the arrangements. Thus, he has demonstrated that ordinary rhythmical respiration—quiet, unconscious breathing—depends on several factors. There is (*a*) an inspiratory mechanism at the level of the striæ acousticae; this he calls the “apneustic centre,” because when this group of nerve-cells is cut off from above, prolonged tonic contraction of the inspiratory muscles ensues (“apneusis”). The level of the striæ acousticae is upper medullary. (*b*) Just below this is a separate expiratory centre (medullary) the existence of which has long been suspected and is now apparently established. (*c*) Both (*a*) and (*b*) are controlled by a higher centre in the upper half of the pons, styled by Lumsden the “pneumotaxic” centre, because it regulates normal quiet breathing. When it is cut off from (*a*) by appropriate section, respiration takes the form of a series of prolonged inspirations, each followed by two or three relatively quick expirations of abnormal type. Lumsden has shown that this cycle repeats itself with great regularity. Evidently, then, the pneumotaxic centre effects normal respiration by inhibiting the activity of the apneustic centre below (behind) it. (*d*) A fourth, “gasping,” centre, situated below (*b*) at the level of the apex of the calamus scriptorius, is regarded by Lumsden as a “relic,” and need not further concern us.

No mention is made by this writer of the position on transverse section of the various groups and tracts the functions of which he has demonstrated, nor is there any allusion to concomitant implication of the face in respect of the activity of the pneumotaxic centre—not that this, perhaps, was to be expected. It is therefore impossible, without further investigation, to say what relation, if any, there may be between the pneumotaxic centre of Lumsden and the postulated co-ordinating centre for the faciorespiratory mechanism referred to above.

3. Our next consideration bears on the influence of voluntary action on the combined respiratory centre in the pontomedullary apparatus. Its automatic activity is set aside *voluntarily* when we deliberately hold our breath, or when we voluntarily pant, cough, yawn, sigh, take deep breaths, etc. Further, its activity is set aside *involuntarily* when we are convulsed with laughter, or when we give way to crying, sobbing, howling. In both the former and the latter case facial movement is involved; we innervate the

facial musculature *voluntarily* for the purposes specified, and the face takes its share in the *involuntary* expression of joy or sorrow.

Thus we get the idea of a *double control* over the faciorespiratory synkinesis: (a) a voluntary control when we choose to inhibit automatic movement, and (b) an involuntary control when that automatic movement is forced to give way to the expression of emotion.

(1) **Voluntary Control.**—The path followed by volitional impulses to facial and respiratory muscles is undoubtedly the familiar corticopontine, corticobulbar, and corticospinal tract. In particular, the geniculate bundle of the pyramidal tract, from the operculum and lower end of the precentral gyrus, via the genu of the internal capsule, conveys these impulses to the appropriate nuclei. As we have seen, voluntary breathing sets aside ordinary breathing, hence we must postulate, on the principle of reciprocal innervation, a synchronous inhibition of the automatic pontobulbar centre. The anatomical course taken by the latter, inhibitory, impulses is less certain, but of their reality there can be no question. It will be remembered that Hughlings Jackson⁵² explained the interesting observation he made on respiratory movement in hemiplegia by the existence of double sets of respiratory fibres passing from the brain in this way.

Lesions, therefore, of the geniculate bundle anywhere in its course—especially if they are bilateral—will impair volitional control over the musculatures concerned in the expression of emotion, with the result that the involuntary action of the same mechanisms will tend to become abnormal. Pseudobulbar paralysis is the disease of the geniculate bundles which, we have already seen, is especially prone to be accompanied by the phenomena of *rire et pleurer spasmodiques*. If the reader will refer again to Case 3 above he will note there was *absolute voluntary paralysis* of face and of respiratory apparatus—hence emotional seizure of the same parts was unchecked, and the patient's existence was one long roar of laughter. An old observation recorded by Magnus,⁵³ in 1837, presents certain analogies to my Case 3, and may be briefly outlined. The patient was a widow of 50 who had had two strokes, with the result that there was *complete* bilateral paralysis of the face and tongue; yet she smiled and laughed, often violently, the paroxysmal laughter ending in a peculiar, grunting sound of which she was ashamed and which she would willingly have suppressed;

it continued, however, even after the movements of laughter had ceased.

It is clear, then, that the more severe the volitional facio-respiratory paralysis, the more exaggerated is involuntary innervation of the same mechanism. In this connexion Monrad-Krohn has shown that emotional innervation is often distinctly exaggerated on the paretic side in hemiplegia, and has proved (by slow-motion cinematography) that emotional movement is actually quicker on the side showing voluntary paresis. On the other hand, for the exhibition of uncontrollable laughter or tears a degree of volitional paresis or paralysis is not quite essential, though it is certainly usual; the involuntary action of a normal laugh may break down normal control; the quivering lip of the child is indicative of a balance between the action of voluntary and involuntary processes which may be tipped over in either direction by a trifle.

(2) **Involuntary Control.**—Careful experiments of W. G. Spencer,⁵⁴ in 1894, determined the existence of four paths from cerebral cortex to respiratory mechanism. Of these, one is undoubtedly the voluntary path just mentioned, from the motor cortex via the genu of the capsule; its stimulation produces, in the ape, a sort of holding-the-breath, or, as Spencer calls it, “over-inspiratory tonus.” Two of the other tracts follow an entirely different course; one is an arresting and the other an accelerating path. The former arises from the under surface of the frontal lobe, the latter from the sensory cortex. Spencer has traced the two throughout their course; they come together towards the middle line at the mesial aspect of the lower optic thalamus, bordering on the third ventricle, and run down, near the midline of the tegmentum, to the medulla. Both are far removed from the voluntary tract for respiratory innervation in the capsule and crus. More exactly, the route followed by the *arresting* path is from a spot on the under surface of the frontal lobe where the olfactory tract runs into the temporosphenoidal lobe, along the olfactory limb of the anterior commissure (where it decussates), by the side of the infundibulum, past the nucleus ruber below and external to the aqueduct in the plane of exit of the third nerve, and so to the medulla. As for *acceleration*, “commencing especially from a point on the convex surface of the cortex within the sensorimotor area, the effect may be followed back through the lenticular nucleus where it borders on the outer and ventral portion of the internal capsule; the strand runs at first externally and then ventrally to the

motor portion of the internal capsule, and so reaches the tegmentum. The lines from the two sides meet in the interpeduncular grey matter at the level of and just behind the plane of the third nerves."

The figures reproducing Spencer's photographs (Figs. 54 and 55) indicate the position of the arresting and accelerating respiratory paths and show their distinction from the voluntary tract for respiratory innervation in the capsule and crus.

I believe it is a feasible speculation that these are the paths for emotional activation of the faciorespiratory mechanism. They are separate from the paths for voluntary control; they come towards the midline in the regio subthalamica and tegmentum;

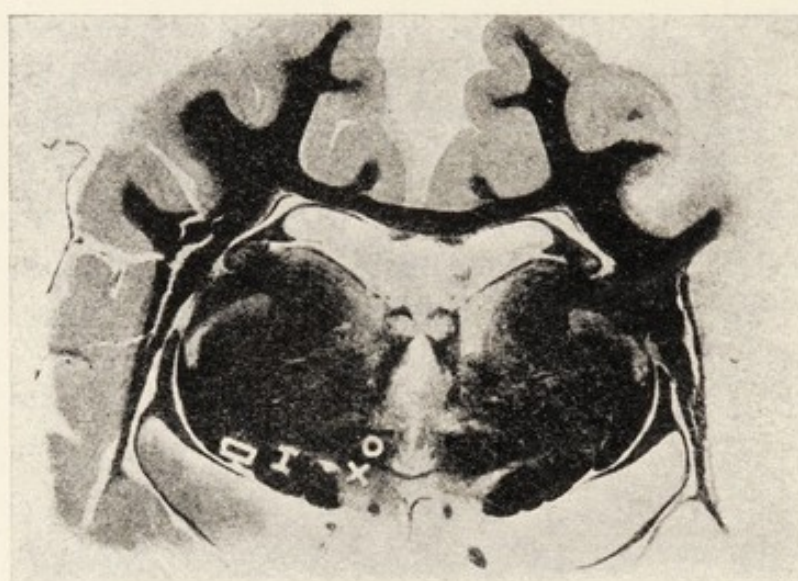


FIG. 54.—(Spencer.)

O = position of respiratory path, stimulation of which produces slowing and arrest.
X = ditto, stimulation causing acceleration. These are extrapyramidal tracts.

stimulation of them produces unvaryingly the phenomena of arrest and acceleration noted above. As far as the respiratory element in involuntary laughing and crying is concerned their appropriate excitation and inhibition will explain the mainly expiratory character of the former and the mainly inspiratory character of the latter.

Clinical proof of the reality of the faciorespiratory involuntary synkinesis, and of the possibility of combined unilateral paralysis of its two components, is furnished by Stromeyer's¹² remarkable case, already mentioned. It remains to ascertain if we have any experimental evidence bearing on the association of the face with involuntary respiratory tracts. In a useful paper entitled "Note on the Physiology of the Basal Ganglia and Midbrain of the Anthro-

poid Ape, especially in reference to the Act of Laughter," Graham Brown ⁵⁵ has given us certain data that bear on our subject. He has demonstrated in the normal animal that tickling in the hollow of the shoulders, armpit, etc., causes the chimpanzee to respond by retraction of the lips, as in smiling, while at the same time the respiration becomes more rapid and slightly vocal. "The sound given is that of 'Ha, Ha, Ha,' but not said as we say it—rather whispered. There can be little doubt that this reaction to tickling is equivalent to the act of laughter." *

The same investigator, working on the exposed surface of the mesencephalon after transection, has found that between the

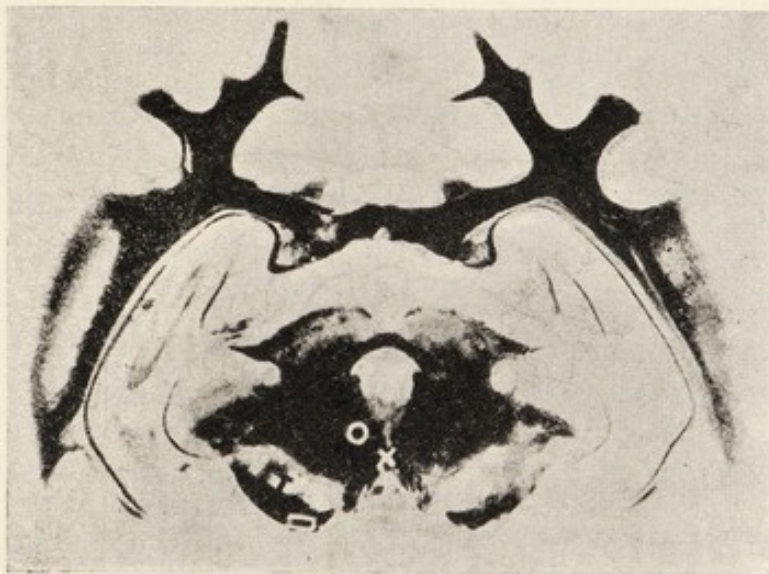


FIG. 55.—(Spencer.)

O and X as in Fig. 54. Note the distinction of these paths from the respiratory tracts in the crus (indicated by \square and $\mathbf{\Gamma}$ in both figures).

internal boundary of the red nucleus and the mid-longitudinal dorsiventral plane of the neuraxis is a small and very strictly circumscribed area, not much more than 1 mm. across, unipolar stimulation of which suddenly changes normal slow, deep, and steady respiration to fast and shallow breathing (Fig. 56). The abdominal muscles of both sides of the body appear actively to contract and relax during the reaction, which stops always with cessation of stimulation. Graham Brown says specifically that "the sound of this breathing was very similar to the 'Ha, Ha, Ha' of the laughing chimpanzee." In a personal communication he states that to the best of his recollection the facial muscles were retracted at the mouth at the same time.

* There is a fine photograph of a "laughing" chimpanzee on page 92 in Crile's book on the Emotions (see Reference 23).

Hence, though there is some slight uncertainty, it would appear that excitation of a specific descending tract in the mesencephalon causes the animal to make both facial and respiratory movements of laughing, apart from corticobulbar pathways. Graham Brown has also found within the extreme caudal end of the optic thalamus two spots, of which stimulation applied to the dorsal one causes very vigorous and "hollow" breathing, while excitation of the ventral spot gives a slowing of respiration.

A comparison of the results obtained by Spencer and Graham Brown shows, as far as mesencephalon and tegmentum are concerned, the existence of resemblances in anatomical position and objective phenomena sufficiently impressive to outweigh such

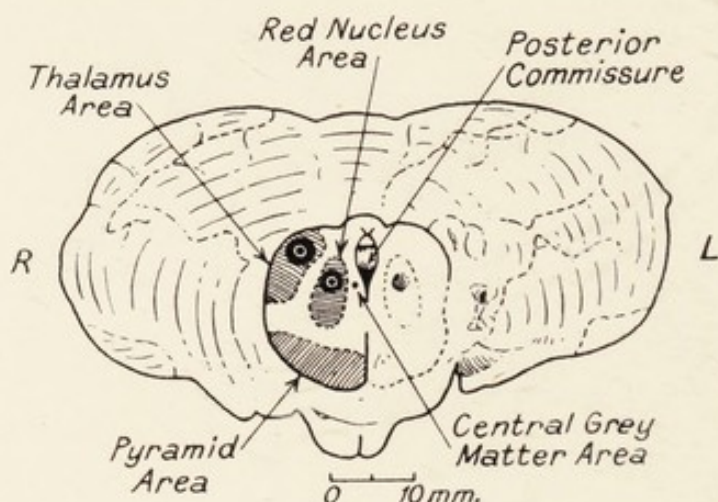


FIG. 56.—(Graham Brown.) Stimulation of the spot indicated as "central grey matter area" produces a modification of the chimpanzee's breathing closely akin to laughing.

discrepancies as still remain. Their correlation with the work of Lumsden is difficult, as already hinted, because of the absence in the latter's experiments of evidence pointing to a particular localization on transverse section at the level of the upper pons.

Our general conclusion may be couched in the following terms: There are corticifugal paths to faciorespiratory centres in pons and medulla that are independent of voluntary cortico-ponto-bulbar tracts to the same nuclei; on excitation they will either arrest or accelerate, i.e. interfere with, normal rhythmic activity of the respiratory centre; the available evidence warrants the speculation that they are the routes taken by emotional impulses to modify the faciorespiratory synkinesis in the direction either of laughter or the reverse. Their exact course remains for further determination; it is perhaps noteworthy that they make their way separately

towards the midline, skirting the lower optic thalamus (in the case of one) and passing by the lower regio subthalamica to the tegmentum, and so to more caudal levels of the neuraxis.

Application to Diseased Conditions

1. Pathological laughing and crying are allowed by lesions of the voluntary paths from the motor areas of the cortex or by any state in which these exercise imperfect control. Laughing and crying then become uncontrollable. Their common appearance in pseudobulbar paralysis is readily understood, because of the usual volitional facial, etc., weakness.

It must, however, be pointed out at once that this is not a complete explanation of the facts. Some cases of bilateral facial weakness of central origin are not particularly prone to develop the exaggerated emotional display of which we are speaking, while it may appear in other diseases in which bilateral voluntary control is not in any way impaired, or not obviously impaired, by the morbid state. In the case of the former it is likely either that the individual is not by constitution particularly emotional, or that voluntary control, though defective, is still adequate, or that, possibly, disease is affecting the activity of the non-voluntary paths as well as of the voluntary. In this connexion Féré's case of *fou rire prodromique*, referred to above, is of interest in view of the cessation of uncontrollable laughter subsequent to development of a severe left hemiplegia. It would have been of value to ascertain the position of the lesion or lesions which thus caused the emotional exhibition to stop.

In the case of the latter—as, indeed, in normal persons—emotional stimuli evidently overwhelm the control mechanism, and we must suppose either an irresistible quality in them, heightened by disease; or a constitutional peculiarity on the part of the individual—disease apart; or defect of cortical control not discoverable in tests for volitional facial innervation; or, perhaps, defect on the afferent side to the cortical emotional centres from which the facio-respiratory paths arise.

2. The reverse condition, unilateral (or bilateral) emotional palsy, is brought about by a lesion of the appropriate involuntary system whose possible course has already been indicated, the voluntary cortico-ponto-bulbar tract being normal. Ex hypothesi, this may occur anywhere from the cortex to at least as low as the pons.

For example, the clinical evidence in my Case 8 is strongly suggestive of a lesion in the mesencephalon (paralysis of upward and downward conjugate movement of the eyes, unilateral Argyll Robertson pupil, hemitremor) under the anterior corpora quadrigemina. The patient showed unilateral, right, emotional facial palsy. Similarly, in Case 9, a tumour of the mesencephalon was in a position to effect unilateral mimic paralysis (see Figs. 50 and 51). The remarkable instance of the same condition recorded by Mills¹⁵ was associated with various other symptoms and signs, and was due to a destructive lesion involving, inter alia, the mesencephalon. Allusion has already been made to the occurrence of this paralysis in cases in which the thalamus, speaking loosely, has been implicated. If the hypothesis advanced in this communication is trustworthy, the condition should not occur in all thalamic cases by any means, but only in those where the lesion is so placed as to engage the tracts specified.

We have seen they do not pass right through the thalamus—at least in the case of the arresting path—but rather skirt it mesially and ventrally. The argument, therefore, opposing Nothnagel's original speculation on the ground of the occurrence of negative thalamic cases loses much of its value. The lesion causing mimic palsy does not occupy the same site as that causing the thalamic syndrome—an explanation which has suggested itself to Gordon Holmes⁵⁶ and others, and which is borne out by the considerations here advanced. The two, however, may doubtless be combined, though no definite instance has as yet come under my notice.

Ex hypothesi, further, mimic paralysis may originate in a cortical lesion, but, so far as I have seen, no such case has yet been recorded.

The possibility of a combination of (1) and (2), viz., the occurrence of facial asymmetry in uncontrollable laughter, is realized in Case 3 above, in which, with highly characteristic *rire spasmodique*, the angle of the mouth was not retracted so well on the left side as on the right, and has been seen in other recorded cases.

It is of particular interest to note the general grouping of the cases of mimic paralysis discussed above (Cases 3, 7, 8, 9, Mills' case) in relation to the distribution of the posterior communicating artery. Beever has shown that this vessel supplies the anterior third of the crista, and all the part between it and the third ventricle at the midline; frequently, too, it irrigates the anterior half of the internal nucleus of the optic thalamus. Now we have seen that

the respiratory tracts discovered by Spencer come together within this area; hence a lesion in the distribution of the posterior communicating artery may be regarded as likely to exteriorize itself, *inter alia*, by unilateral mimic paralysis. The explanation given by Beevor for the phenomena of his striking case rests on a different interpretation from my own; he thinks Spencer's respiratory arresting and accelerating paths are those of *volitional* control, and that their involvement caused the paralysis of voluntary respiration in his patient. With this view I am not in accord, as will have been seen; I believe, however, their partial implication explains the incomplete mimic paralysis which his case certainly exhibited.

From the argument here advanced it will be gathered that any hypothesis for the placing of actual mimetic *centres* in the thalamus is unnecessary. The thalamus cannot be more than a link in the chain. Afferent paths for appropriate impulses from eye, ear, skin (tickling), etc., lead through the thalamus to the cortex. The laughter-producing stimulus is cortically appreciated, and its expression through the involuntary faciorespiratory mechanism is mediated, *ex hypothesi*, by the efferent arresting or accelerating tracts already described. The arresting tract in its descent skirts, or perhaps runs through, the lower mesial margin of the thalamus—the palæothalamus, be it noted, which borders on the third ventricle, and which, according to Tilney and Riley,⁵⁷ “seems to be invested with a functional responsibility related to the development of the emotions and the emotive expressions.” On the afferent side neuronic systems are relayed in the thalamus, but it is not certain if the same obtains on the efferent side, hence the possibility of a short-circuit from sensory to motor path in that ganglion itself remains undecided. Whether such a short-circuit would explain the explosive character of spasmodic laughing and crying, the exaggerated response to trifling emotional stimuli, is equally uncertain. The argument from pseudobulbar cases is that some defect of volitional faciorespiratory control heightens the facility of the explosive phenomena, yet, as we have seen, it is not always *per se* sufficient. It is conceivable, therefore, as some have maintained, that failure of corticothalamic inhibition is responsible for undue liveliness of the thalamus, and for the exhibition of involuntary emotional exaggeration.

For myself, however, I am of the opinion there is more to be said for the participation of the cortex in the production of abnormal

emotional activity. We cannot take it that the cortical origins of the arresting and accelerating respiratory tracts of Spencer are physiologically, though anatomically, separate, and we may ask—using Mills' expression—where is the rendezvous? In an ingeniously developed argument, that veteran neurologist^{58, 59} contends that in the right hemisphere mainly, in the midfrontal region, are centres for the representation of movements especially concerned with the expression of emotion. He gives the term "movement" a broad significance, as applying both to skeletal and to visceral, vascular, and secretory activity. On the other hand, Bianchi,⁶⁰ whose claim to speak with authority also is acknowledged, declares that "to maintain that the frontal lobe plays a part in the essence and mechanism of the emotions . . . is a bold hypothesis in which there is a good deal of mere conjecture and certainly no basis of proof."

Be all this as it may, and however much in the matter is still obscure, our facts have led us to suggest that there are corticifugal paths for the expression of the emotions via the faciorespiratory apparatus, distinct from those for voluntary innervation of the same nuclei, and as a necessary corollary we presume the existence of a cortical nodal point co-ordinating them. Its situation is at present indeterminate, yet it is likely to have some definite position. In this connexion I echo with approval the words of Mills, who declares he is not one of those who believe that the problem of emotion, or of any other great mental process, is to be explained by regarding it in some vague way as a complex expression of the action of the cerebral cortex as a whole.

There is clinicopathological and experimental evidence suggesting that non-volitional control over the normal automatic activity of the faciorespiratory mechanism is exercised from the cortex by routes that pass separately downwards to come together towards the midline in the regio subthalamica and tegmentum.

It is not certain that these actually pass through the thalamus in man, though it is understandable that some thalamic lesions may be so placed in that ganglion as to interfere with them as a vicinity effect.

We have no information as yet to show these paths are interrupted by a thalamic relay nor is it known that emotional impulses can pass from sensory to motor side at this level; it is, perhaps, a possibility.

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CHAPTER XIII

DYSÆSTHESIÆ AND THEIR NEURAL CORRELATES *

Preliminary Considerations. Levels of Lesions associated with Dysæsthesiæ : Peripheral; Spinal; Bulbar; Thalamic; Cortical. Pathogenesis: Excitation and Inhibition. Dual Mechanisms in the Sensory System. Criticism of a Jacksonian Theory. Corticothalamic Inhibition. The Neurovascular Factor in Pathogenesis. Explanation of Hygriæ Dysæsthesiæ.

The clinician is frequently confronted with cases a prominent feature of which is the occurrence of "subjective" or "spontaneous" sensations of one or other modality. Terminology here is not a little confusing, for all sensations are subjective, being experienced in consciousness by a subject, whether they arise from extrinsic stimulation or not. Further, use of the term "spontaneous" is unsatisfactory, since excitation by some form of stimulus, extrinsic or intrinsic, however elusive, trifling or untraceable it be, must be postulated if sensation is to come into existence. It has been customary to include under the expressions "paræsthesiæ" or "dysæsthesiæ" abnormalities of sensation appearing to develop spontaneously, but agreement has not been reached as regards the precise meaning of the terms. "Paræsthesia," for example, signifies, according to the French school, any disorder of sensibility which cannot be classed either as an "anæsthesia" or a "hyperæsthesia." The latter, however, is as a rule diagnosed only by objective exploration of a given part, as is the existence of "anæsthesia"; and while it is true the patient often enough complains of a sensation of "numbness" or "deadness" in an area shown objectively to be distinguished by loss of appreciation of cutaneous stimuli, the clinician rarely if ever employs the term "anæsthesia" for this subjective sensory condition. Moreover, objective anæsthesia may be unassociated in consciousness with any subjective phenomenon of the above class.

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In view of these and other somewhat conflicting interpretations, it seems preferable to utilize the word "dysæsthesiæ" comprehensively to include all apparently spontaneous phenomena described as "burning," "coldness," "wetness," "numbness," "deadness," "tingling," "pricking," "pins and needles," "sensation of something moving," etc. There is no end to the catalogue, one might think, especially if we are to incorporate picturesque attempts on the part of the hypersensitive neurasthenic to give expression to his cephalic discomfort by such similes as: "I feel as if my brains were being stirred about with a red-hot poker." I shall, however, confine my discussion to sensations arising in connexion with organic lesions of the somatic sensory system, for which a classification more or less in accordance with the usual divisions of sensory experience is possible, as follows:

- (1) Tactile dysæsthesiæ.
- (2) Painful dysæsthesiæ.
- (3) Thermal dysæsthesiæ.
- (4) Complex dysæsthesiæ (e.g., wetness).
- (5) Dysæsthesiæ with sensation of movement.

Preliminary Considerations

Certain preliminary considerations may pertinently be dealt with at this point.

(1) If the phenomena which here concern us may be thought of as "subjective sensations," in contradistinction to "objective sensations" produced by known objective excitations, the fact remains that they are not to be regarded as differing in any subjective respect from the latter; the experiencer is aware of a sensation of coldness, for instance, comparable to that effected by application of a cold substance to the skin; only because of the apparent absence of extrinsic stimuli is its description as "subjective" justifiable, since, as we shall see, its objective causation by some abnormal physiological factor can, in many cases at least, be fairly established, and in others must be assumed.

(2) Sensations are psychical things, arising in association with physiological processes in the nervous system. In the course of this study evidence will be adduced to prove that dysæsthesiæ appear in consciousness as a result of lesions at differing physiological levels—peripheral, spinal, bulbar, thalamic, cortical; but we must bear in mind that sensations, however variously derived from the physiological viewpoint, are constituted uniquely by

changes in consciousness. Since the term "receptor" is in common use for a sensory end-organ, Parsons¹ has recently suggested that ambiguity would be removed were "reception" and "recept" to be employed psychologically for the act of sensing and the object sensed. It would then be easier to distinguish between sensation and perception, for the latter, the act of perceiving, consists in the integrating of sensations ("recepts"). Even when the sensation is simple it does not constitute the fundamental psychological unit—this is the percept, and there is no such thing as a pure or isolated sensation. When, therefore, we shall have occasion to speak of the "subjective sensations" or *dysæsthesiæ* of our cases, we cannot stop short at any point as though these existed by themselves; we must consider also the emergence of the sensations in a perceiving consciousness, their possessing affective tones and cognitive accompaniments.

We cannot know the sensations of our patients except by their description of them, which of course is a matter of speech. Elementary as this remark is, it has a distinct bearing on at least one aspect of the problem; for it implies that the way must be open between the sensation-experiencing mechanisms and the cortical speech-mechanism, and this is of significance, as we shall see subsequently, in connexion with the subjective abnormalities of the thalamic syndrome.

(3) It follows from the above consideration that *dysæsthesiæ* exhibit the usual variations in degree, extent, duration, of all sensory experiences; they are modifiable by attention, and by other factors of the psychical series. At the same time, their essential crudeness, relative simplicity and primitiveness must be emphasized. The presentations are of recepts which can be analysed and discriminated one from another, but in a given case as a rule only one form is present at a time, combinations of the painful plus thermal type, or tactile plus thermal type, being on the whole less frequent. As remarked already, in a general sense *dysæsthesiæ* are considered to be "spontaneous," but clinically we find numerous illustrations of the fact that they may be influenced by extrinsic agents, such as cold or warm currents of air, the contact of substances (e.g., water of differing temperatures) on the parts affected, and so forth. Further, the clinician often observes cases where *dysæsthesiæ* do not seem to arise in consciousness unless and until some such extrinsic excitation occurs, as when the patient declares, for instance, that the wind blowing on his face awakens such-and-

such an unpleasant sensation, or when the causalgic sufferer is in comparative comfort until an extrinsic stimulus (it may be of another order than the cutaneous sensory kind) arouses unbelievable pain. It would be rather undesirable to exclude such cases from the "spontaneous" category, for the good reason that excitation frequently only accentuates or aggravates an already existing if faint dysæsthesia. On the other hand, there appear to be equally numerous cases in which subjective sensations remain uninfluenced by extrinsic stimulation.

(4) Since we shall be dealing with painful dysæsthesiæ and spontaneous "central" pains, a brief preliminary excursus on the nature of pain will not be out of place.

While sensation and perception are distinct processes—considered psychologically—the former cannot occur independently of the latter. No sooner is a sensation felt than the localization of the point stimulated and the interpretation of the particular variety of sensory excitation follow as associative reactions; a cognitive element is inevitably added, and an affective element as well; indeed, the sensation may evoke symbolic representations of another kind (e.g., verbal) and may issue in motor reactions. Perceptive function bridges the gap immediately between the purely sensory and the intellectual. When we speak, as we constantly do, of "sensations of pain," or of the "sense of pain," this consideration must not be lost sight of; testing clinically for pain appreciation, thinking physiologically of pain-spots, pain-fibres, pain-centres, we are apt to overlook the fact of its being something more than a pure sensation. As Piéron² says, "une sensation de 'douleur' est aussi en réalité un complexe de sensation et de réaction affective," and the receptive systems concerned therewith only enter into thought as a result of "répercussions affectives." He makes an exception, however, of pricking, holding that the character of the sensation can be fairly well discriminated and isolated from the "répercussion douloureuse," unless this is exaggerated. Some recent writers, e.g., Foerster,³ prefer to speak of a *feeling* and not a *sensation* of pain (*Schmerzgefühl* as opposed to *Schmerzempfindung*). The association of pain feeling with sensations of another kind is well recognized. Thus a pin-prick awakens a "sensation" of touch-pressure of a point plus a "feeling" of pain; the application of extreme hot or cold stimuli arouses the specific "sensation" plus a "feeling" of pain (*Wärmeschmerz*, *Kälteschmerz*), which by introspection can be felt to come later and to last longer than the

hot or cold sensation. Yet under other conditions, particularly in pathological states, pain exists by itself and cannot be considered other than a sensation loaded with a specific affective tone of unpleasantness. This is undoubtedly the case with the "central pains" of nervous disease and with other varieties of neuralgia. Again, in certain diseased states other sensory systems, those for instance concerned with the appreciation of tactile and thermal excitations, may be incapable of conduction, while painful excitations can still be experienced in consciousness; in cases of pure lesions of the dorsal columns of the cord a stroking touch on the skin can cause unbearable pain—a reaction included by Foerster under the useful term "hyperpathia," which must be carefully distinguished from hyperalgesia.

From the clinical standpoint, if the above-mentioned amplifications are borne in mind, it remains convenient to speak of sensations of pain and of physiological systems concerned with the conveyance of painful impressions, as well as of cerebral centres or zones the integrity of which is essential for the appreciation of pain in consciousness.

Levels of Lesions associated with Dysæsthesiæ

(1) PERIPHERAL LEVEL

With lesions involving sensory protoneurones different forms of dysæsthesia are prone to make their appearance. These are observable perhaps as well after alcoholic injection of the trigeminal nerve or Gasserian ganglion as anywhere else; my first illustrations will be from trigeminal cases.

CASE 1.—A. C. M., female, age 57. Has suffered from left facial neuralgia for three years. In January, 1927, I injected the inferior division (the regional distribution of the pain) at the foramen ovale. There has been no neuralgic paroxysm since, and the corresponding cutaneous area remains anæsthetic. But dysæsthesiæ abound, as the following excerpt from a letter of March 28 reveals: "My face still gives me a lot of trouble. I cannot describe the various feelings in it. Sometimes it is very, very *cold*; then again it will *burn* very much; it is burning a great deal to-night and is almost unbearable. . . . Sometimes my lower lip feels as if it is *swelling* so much that it will burst. . . . Again it seems as though there are threads *pulling* from my temple to various parts of the side of the face."

In this lower division thermal, tactile and painful stimuli are no longer appreciated.

CASE 2.—M. A. F., female, age 58. Has suffered from right trigeminal neuralgia for eight years, mainly in the middle and lower divisions. The patient complains of a constant feeling as though the cheek were *wet*, and she frequently puts her hand up to wipe it, but it is quite dry. I could not satisfy myself of objective impairment of any form of sensibility. After a gasserectomy the *wet feeling* disappeared.

These two cases have been selected with a view to furnishing a comparison and contrast; in the second the hygric dysæsthesiæ were spontaneous, unassociated with recognizable objective sensory change, and vanished after operation on the ganglion; in the first thermal and other dysæsthesiæ only occurred after interference by injection, and developed in an area objectively anæsthetic, thus approximating to an anæsthesia dolorosa.

Other illustrations of dysæsthesiæ at the periphery can be taken from experience of *causalgia*, the "burning pain" described originally by Weir Mitchell, Morehouse and Keen⁴ in 1864, during the American Civil War, in a slender volume devoted to injuries of nerves. From this *locus classicus* a few passages of significance will be quoted.

"It is a form of suffering as yet undescribed,* and so frequent and terrible as to demand from us the fullest description."

"We have some doubt whether this form of pain ever originates at the moment of the wounding; but we have been so informed as regards two or three cases."

"The seat of burning pain is very various; but it never attacks the trunk, rarely the arm or thigh, and not often the forearm or leg. Its favourite site is the foot or hand."

"Its intensity varies from the most trivial burning to a state of torture which can hardly be credited."

"The part itself is not alone subject to an intense burning sensation, but becomes exquisitely hyperæsthetic."

"Most of the bad cases keep the hand constantly wet, finding relief in the moisture rather than in the coolness of the application."

"It appears quite certain that in cases of glossy skin burning always exists. It is also certain that it may exist without association with diseased skin."

"The temperature of the burning part we have always found to be higher than that . . . of corresponding points on the other half of the body."

"If the burning were a referred sensation, it would sometimes be met with

* For the sake of accuracy in neurological history and because of its value, I think it right to point out that a case of *causalgia* conforming to the descriptions of half a century later was reported by Alexander Denmark, under the title "An example of symptoms resembling *tic douloureux*, produced by a wound in the radial nerve," in the *Medico-Chirurgical Transactions*, December, 1813, vol. iv, p. 48. The case was that of a soldier wounded at the storming of Badajos, April 6, 1812.

in cases of complete division of nerves, and, therefore, in parts devoid of tactile sensation. But we have encountered no such cases; and, on the other hand, the burning pain is often accompanied with hyperæsthesia, while motion and touch may remain unaltered."

The important and peculiar features of causalgia may thus be tabulated: (1) It occurs with some lesions of peripheral nerves only, and in some nerves (e.g., the median) more regularly than in others. (2) It does not develop if the nerve is cut across, and is often at a maximum where physiological discontinuity is minimal. (3) It is aggravated by warm or dry applications, and relieved by wet and to a less extent by cold. (4) It is accompanied by phenomena of a different order from those of pure sensation, viz., local raising of skin temperature (not constant), local sweating, local alteration in skin colour (intensification of pinkness or redness of skin, even to purple, sometimes in a rather blotchy way); in other words, phenomena of a vascular and neurosympathetic kind are superadded. A further feature of no little interest is the excitation or accentuation of the pain by stimuli of an affective kind, and by those belonging to other sensory systems—so-called *alloparalgia* or *synæsthesalgia*. The vascular and neurosympathetic accompaniments point significantly to the conclusion that part at least of this peculiar sensory syndrome is attributable to implication of fibres not running entirely with ordinary nerve-trunk fasciculi. In fact, the question arises whether under certain conditions the appreciation of pain is independent of anatomical paths of the peripheral nervous system proper, and further evidence in this respect will be examined more closely at a later stage.

From a large collection two illustrative examples will be chosen.

CASE 3.—*Causalgia of the radial branch of the left musculospiral*.—Private McC. was wounded at Loos on September 25, 1915. A bullet entered the left arm just above the elbow-joint and passed out obliquely in a downward direction. When hit, he immediately felt pain down the left hand and fingers, and in the course of a few weeks this became unbelievably acute. It appeared instantly in paroxysms of burning pain on the most trifling provocation, e.g., when the patient in the next bed turned, when music was played, even when his name was called out by the masseur at the ward door.

Operation showed the sheath of the radial nerve to have fragments of metal and of bone sticking into it. After it was freely opened and stripped from the nerve, to its junction with the musculospiral, considerable relief was obtained; the causalgia largely disappeared, yet not entirely, and emotional stimuli similarly ceased to be algophoric.

CASE 4.—*Causalgia of the internal plantar branch of the internal popliteal.*
 —Private M. was wounded at Loos on September 26, 1915. He was shot through the right leg just above the knee and immediately felt acute pain on the inner side of the sole of the foot. So convinced was he of a wound there that he sat down and took off his boot and sock, but saw nothing. A few minutes later blood began to ooze through his trousers above the knee. In the course of some weeks the pain, which had never ceased, became truly agonizing. The patient sat in bed holding his foot with his hands, pressing it with wet cloths; light touch and pressure caused intolerable burning pain, but firm pressure rather relieved it. A warm or a dry application was insufferable. He used to pour with sweat when the pain was intense. Local phenomena of sweating and altered coloration were always in evidence. Insignificant stimuli (a cough, a patient passing the bed, a laugh, touches on the opposite leg, a rustling newspaper) produced unbearable torture. At operation, just below the branch to the outer head of the gastrocnemius a definite spindle-shaped swelling was found on the internal popliteal, and the minute vessels on its surface were hyperæmic. The sheath was stripped off and the nerve freed; the spindle-like swelling was incised longitudinally in various places and the nerve surrounded by fat.

Immediately after the operation the spontaneous causalgia ceased to some extent, though not entirely; and emotional stimuli failed to exert their previous influence.

Testing for objective changes in sensibility is usually a matter of some difficulty in cases of causalgia, as can be realized best by those who have had occasion to try; generally speaking, tactile, thermal, and painful excitations are appreciated, yet the sensation is not readily separated from its affective accompaniment, since the intensity of the burning pain submerges other qualities of sensation in consciousness. I have not found more than a hypalgesia or hypæsthesia in most instances, while many exhibit an actual hyperalgesia and hyperpathia, the threshold to painful stimuli being lowered, and the response being altogether disproportionate to the stimulus.

The causalgic dysæsthesia is not known to occur in its full form, with vascular and neurosympathetic concomitants, except at the peripheral level; it never develops with lesions of the dorsal roots—a significant observation.

Attention must next be directed to the association of dysæsthesia with other peripheral cases distinguished by the presence of definite symptoms and signs of a *vascular* and *vasomotor* kind. I may mention the large group of acroparæsthesia, characterized by the development of painful and other dysæsthesiæ, nearly always in elderly women of a hard-working class, who commonly exhibit indi-

cations of commencing arteriopathic change. This is an out-patient type par excellence, and one which is clear-cut clinically. It has always appeared to me feasible to link the occurrence of the unpleasant dysæsthesiæ largely at night or in the morning with insufficient blood-supply to the vasa nervorum, a meiopragia or relative ischæmia following on lowered vascular tone during the night hours in minute vessels which are also degenerating.

Other clinical groups, those of acro-asphyxia, acrodynia, etc., and, more generally, those of the Raynaud and sclerodermia categories, have a similar tendency to exhibit dysæsthesiæ often of a chronic and rather intractable kind. Nor should those which accompany intermittent claudication be omitted in this connexion. In all of these examples the interrelation of *subjective sensory disorder* and *altered vascular states* is unmistakable. A recent case of a peripheral syndrome of the Raynaud-sclerodermia class has afforded a good chance of demonstrating the close connexion of the sensory system with the vascular system.

CASE 5.—Miss E. C., age 37, a patient of Dr. C. W. Somerville, of Bonnyrigg. For no less than twenty years the left hand has been the seat of chronic changes involving all the tissues from skin to bone. Some eighteen months ago the right hand, previously normal, began to exhibit similar alterations, which now are as advanced as in the other. In summer the hands are thin and dead-like; in winter they tend to swell up to some extent. The skin of both is shiny, glossy, often discoloured to a bluish-violet hue, and sometimes additionally disfigured by areas of blister-formation. The ends of the fingers, particularly those of the ulnar side, exhibit a panatophy, rendering them hard and claw-like. The skin temperature of the hands is much below that of the rest of the arms. At the wrist on both sides the radial pulse is very faintly palpable. The patient complains of *absolutely unbearable pain* in both hands, resembling that of causalgia, in that it is aggravated by sensory stimuli of another order. She prefers to keep them cold, since warmth causes intenser suffering, and to let them depend.

Objective examination reveals extreme hyperæsthesia, also as in many causalgic cases, to touch contacts. Cotton-wool is everywhere appreciated, and touches accurately localized. Painful and thermal stimuli are normally appreciated; the former, and hot stimuli, exaggerate the continual pain, while cold applications do not.

In view of the general resemblance of the symptomatic picture to that of the causalgia syndrome, I suggested a periarterial sympathectomy on the left side, and this was agreed to. On January 17, 1927, the patient was operated on in Edinburgh by Mr. Quarry Wood.

The *brachial artery* was duly exposed at its upper end in the left arm and was recognized only with considerable difficulty, as it was extremely small, no thicker than an ordinary match. Its pulsations were very slight, indeed

almost imperceptible. The operator decided that to attempt stripping the periarterial plexus was impracticable; instead, following the technique of Sampson Handley, he injected a drop or two of 95 per cent. alcohol into the arterial wall, which immediately turned white over a distance of about a quarter of an inch, or rather more, while pulsation ceased at the wrist, though it returned some hours later. The wound healed by first intention.

Soon after the hand felt warm for the first time, and at intervals since it has been warm for a period of many minutes, but it never keeps warm, always reverting to its previous cold state. The pain was at first distinctly "different," being less intense and much less continuous. When I re-examined the patient, however, on February 11, it was just as agonizing as before, and no objective modification of the results of the original sensory tests could be found.

From this puzzling case certain deductions can, it seems to me, be justifiably drawn. The normal responses (normal, that is, apart from the hyperæsthesia and hyperpathia) indicate anatomical integrity and physiological conductivity in the nerves of the arm; the concomitant trophic changes, skin discoloration, low surface temperature, etc., suggest persistent vasomotor alteration in the form of angiospasm, and with these the remarkable reduction in calibre of the brachial artery must, I think, be associated. That alcoholic injection into the artery sheath "changed" the nature of the pain for the time, and also exercised a modifying if temporary effect on the coldness of the extremity, is of neurological interest. I take this case to suggest that under certain conditions intense pain may be experienced in a limb, derived not from a morbid process in peripheral nerves but from changes in periarterial plexuses of non-medullated nerve-fibres and accompanied, it may be, by vasomotor disorder. Such an origin for the pain seems feasible, though the difficulties in the way of the proposed operation prevented fuller proof of the conjecture, and the case, at present, remains incomplete.

Valuable corroboration of the general contention, however, is afforded by Tinel,⁵ who in cases of causalgia from wounds of the wrist and forearm was able to "*constater l'existence de troubles dans tout le domaine du sympathique cervical, avec rétrécissement et vaso-constriction de toute l'artère humérale, dont le calibre ne dépassait pas 2 ou 3 millimètres et dont les battements étaient à peu près nuls*" (italics mine). Analogous cases have been published by Brüning.⁶ The similarity of the vascular and sympathetic condition to that of Case 5 is very impressive.

(2) SPINAL LEVEL

The appearance of different dysæsthesiæ at the spinal level is a commonplace in such conditions as disseminated sclerosis, subacute combined degeneration, tabes dorsalis, and syringomyelia, structural changes implicating either the sensory protoneurones proximal to their spinal ganglionic centres, or the deuteronurones of the cord itself. Conformably with what is known of the re-arrangement of sensory systems in the cord, greater dissociation, i.e., more definite separation, of one type of dysæsthesia from another is to be expected, and is found perhaps in the majority.

Without alluding to all the varieties encountered, I may select painful and thermal types, as well as those of a mixed character.

The occurrence of spontaneous or *central pains* and painful dysæsthesiæ in organic spinal disease is now well recognized. They have been studied by Holmes⁷ in cases of gunshot wounds of the cord, and have been noted in an occasional case of disseminated sclerosis, in hæmatomyelia, intramedullary tumours and central gliosis, and in syringomyelia. The mechanism of production of these spinal pains falls to be considered in a later section, but I may here point out that they are different from causalgia, and that, in further contradistinction from peripheral cases, they rarely appear in anæsthetic areas—that is to say, the phenomenon of anæsthesia dolorosa often characterizes peripheral and radicular cases, but seldom purely spinal disease. An important case exhibiting this peculiar combination, however, will be described immediately (see below, Case 9).

Thermal dysæsthesiæ in spinal lesions have engaged my attention for years. Along with the late Dr. P. W. Saunders, I collected a number of such cases, but my material has not been published. An impetus to this research was provided by Hughlings Jackson,⁸ who, in the last "Neurological Fragment" he wrote (1909), expressed the opinion that in some cases of continual over-coldness ("subjective sensation" of cold) the patient might be found to exhibit inability to appreciate hot substances objectively in the same cutaneous area. I was able to give Jackson a reference to a case with this double symptomatology recorded by Sir Charles Bell.⁹

CASE 6 (Bell).—"A man who had lost this sense [sense of temperature] in his right hand, but retained muscular power, lifted the cover of a pan, which from falling into the fire was burning hot, and deliberately replaced it, without being conscious of the heat; the effect, however,

was that the skin of the palm and fingers was destroyed. The same man had a continual sense of *coldness* in the affected arm, which actual cold did not aggravate, nor heat in any degree assuage."

Absence of further details renders diagnosis somewhat problematical, yet it is distinctly possible the case was one of syringomyelia. A more definite, personally observed, instance of this disease may now be quoted in support of Jackson's speculation.

CASE 7.—M. B., male, age 44. Diagnosis, syringomyelia. The patient complains of a constant feeling of *icy coldness* over the whole of the right arm and over the right side of the trunk from the line of the neck and jaw to below the level of the umbilicus. In this region there is loss of appreciation of painful stimuli, and loss also of the appreciation of heat in any degree, whereas cold contacts are usually (though admittedly not always) recognized.

Another spinal case, one of *tabes dorsalis*, illustrates the same double symptomatology.

CASE 8.—A. H., male, age 34. The complaint in this instance is one of continual *icy cold* feelings all over the legs and the perineum. "I feel as if I were sitting on a block of ice." "I feel as if a cold draught were blowing over my legs, just as though I had no clothes on." On objective examination the skin temperature of the parts affected is not materially altered from that of normal areas. Loss to pin-prick is complete in the segmental skin areas corresponding to the spinal division from D 12 to S 5, and, in addition, over the whole of this region there is complete loss of thermal sensibility, whereas cold stimuli are everywhere appreciated, although the patient declares they are not so cold as over normal areas.

Thus I am able to adduce clinical illustrations yielding support to Hughlings Jackson's generalization, but I hasten to point out that other clinical cases do not favour it. The exceptions are of two classes: (1) In some cases a subjective thermal dysæsthesia exists with loss of all thermal appreciation; (2) in others it is present though no objective sensory change can be elicited. As an instance of the former of these two groups the following case may be cited.

CASE 9.—A. C., male, age 73. The patient complains of: (1) A constant *burning* feeling over the right side of the abdomen, and to a less extent over the whole of the right leg; (2) *numbness* over that leg; (3) extreme *tenderness* ("like an electric shock") to light touch-contacts over the same areas.

The results of repeated tests may be summarized as follows:

(1) *Touch*.—Over the right leg and up to the level of D 8 cotton-wool touches are much less plain than on the other limb, and several stimuli

are missed. Stroking with a wisp of wool often causes a hyperpathic response scarcely to be distinguished from pain.

(2) *Pain*.—From the level of D 10 downwards light jabs with a pin are nowhere felt as sharply as on the left. They are less “pointed”; “not so like a pin.” Nevertheless they, especially if somewhat harder, provoke more pain than on the other side, “as though the right were more tender.” In other words, the response on the right partakes of the nature of hyperpathia.

(3) *Temperature*.—The patient is unable to distinguish directly either heat or cold over the region from D 8 down, i.e., there is complete objective thermal anaesthesia in this area. But indirectly a considerable distinction can be drawn as between the two modalities. Thus, when touched on the right side with a cold tuning-fork or cold hand, he jumps, for cold produces a sensation of extreme painful tenderness. Warm or hot contacts, on the other hand, are felt as touch (“like wool”). Hence he can differentiate between heat and cold by noting that a hot test-tube produces a “touch” sensation, and a cold tube a “tender” sensation.

(4) The objective *difference between the skin temperature* of the right and left abdomen and legs is at once apparent to the hand of the observer, as it is to the patient himself. Repeated examination with a skin thermometer reveals a difference of nearly 2° F. on an average between the two sides, more noticeable on the abdomen than the lower limbs.

(5) Finally, there is a constant *difference between the systolic blood-pressures* in the two legs, amounting to as much as 20 mm. Average readings are as follows: Right leg, 105 to 110 mm.; left leg, 125 to 128 mm.; (right arm, 125 to 130 mm.).

Scrutiny of this instructive case allows some conclusions of considerable importance for our subject. The symptoms point to the existence of a vascular lesion in the cord, possibly of the nature of a localized myelomalacia, with its upper limit at the level of the seventh or eighth thoracic segment and occupying mainly, as it seems to me, the dorsal horns over a considerable extent in a downward direction. In favour of this localization is the fact that the right “ano-genital flap” does not escape; evidence (both experimental and clinicopathological) favours an extrafunicular conduction of pain impulses from this cutaneous area by what Foerster³ calls “a continuum of numerous short chain neurones” in the spinal grey matter of the same side. The absence of motor symptoms, changes in reflectivity, or sensory alterations on the left side negatives anything like a complete Brown-Séquard lesion. There may be some slight implication of touch paths passing in by the dorsal horn entry-zone to the dorsal funiculi, but the total thermanaesthesia and the analgesia are in favour of a dorsal horn lesion mainly. The pain over-reaction, or hyperpathia, can possibly be taken to indicate

that this dorsal-horn conduction is not completely interrupted (but see below).

Of no less interest and difficulty are the two phenomena of local rise of skin temperature with subjective sensation of burning, and local reduction of systolic blood-pressure. To account for these unusual features taxes our knowledge of the central (spinal) representation of the sympathetic mechanisms controlling vascular calibre. It is clear in this case there must be a local vasodilatation segmentally set in action, which conjecturally is connected with the intermediolateral tract and centres in the lateral horns of grey matter on the right side of the cord. For if this speculation be impermissible we are faced with the problem of two separate lesions for the symptom-complex, viz., one presumably spinal, and one in the sympathetic side-chain of ganglia; the likelihood of this latter view seems very remote.

If the lesion is considered to be not right spinal but left thalamic, the chief difficulty remains, viz., how to account for the local vasodilatation, reduction of blood-pressure, and rise of temperature. I have never seen these peculiar phenomena in any typical case of the thalamic syndrome.

Be this as it may, the case is cited in some detail to prove that a subjective sensation of a thermal class, a dysæsthesia of *burning*, can accompany complete *loss of thermal appreciation* objectively, and it therefore does not accord with Hughlings Jackson's hypothesis already outlined.

The second class of exceptions to Jackson's speculation is furnished by cases in which thermal dysæsthesiæ are unaccompanied by objective sensory changes, of which the following case is an illustration.

CASE 10.—W. A., male, age 57. For about one year the patient has been aware of a feeling of *intense coldness* over the lower part of the abdomen, buttocks, lower part of the back and upper parts of the thighs. The severity of this *icy coldness* prevents his continuing at his work. He wraps his genitalia in cotton-wool and swathes his mid-parts with it, to no avail. The only other symptom is some degree of defective vesical control, especially at night. He contracted venereal disease a number of years ago.

Objective examination of the regions affected fails to elicit any change in thermal appreciation, which is everywhere normal (both modes), or any change in painful or tactile sensibility. The sole alteration is a definite diminution of the sense of vibration in both lower extremities. Reflectivity is unaltered. A Wassermann test in the blood gives a positive reaction.

Although the cutaneous area of the cold dysæsthesia does not, perhaps, conform entirely to a segmental distribution, seeing that while it involves lower sacral zones it omits lower but affects upper lumbar regions, nevertheless certain little signs point to an organic basis for the affection, and the absence of objective thermal change is again, therefore, not in accordance with Jackson's views.

As a final variety of spinal dysæsthesiæ I shall refer to the complex feeling of *wetness*.

CASE 11.—C. M., male, age 43. Diagnosis, disseminated sclerosis.

For a number of years the patient has complained of a persistent *sensation of wetness* in the legs, like *trickles of water* passing down to the heels. So realistic is this feeling that, convinced of the wetness of the skin of his calves, he has often rubbed it with his hand, only to find it normally dry.

Objective examination, repeatedly conducted, has failed to discover any change in any form of cutaneous or deep sensibility in the parts involved, with the single exception that tactile contacts are not invariably appreciated with normal acuity over the areas where the dysæsthesia is localized.

In this clinical instance, it is true, the element of "warm" or "cold" is not added to the "wetness," yet the sensation passes beyond one of mere formication, of "something moving," since the "wet" component definitely enters therein. The usual symptoms and signs of disseminated sclerosis of a mixed cerebrospinal class are characteristically present.

(3) BULBAR LEVEL

At the levels of medulla, pons, mesencephalon and regio sub-thalamica organic lesions implicating sensory paths commonly are accompanied by one or other kind of dysæsthesia; every variety is known and has often been recorded. In view of the anatomical dissociation of afferent fibre-groups in the fillets corresponding to the forms of sensory experience (as also at spinal levels, already mentioned), the clinician will frequently find the dysæsthesiæ to be peculiarly distinct and uncomplicated. Lesions of the posterior inferior cerebellar artery, as is well recognized, are especially calculated to offer the combination of subjective sensations with objective sensory change, since they involve the *formatio reticularis* with its ascending spinothalamic paths, as well as the descending root of the fifth and the issuing quintothalamic tracts. Anywhere in the brain-stem below the actual thalamic level spontaneous pains

and dysæsthesiæ of the usual kinds can arise as a sequel to structural disease, and choice of material is almost embarrassing.

My former clinical assistant and house physician, Dr. W. G. Wyllie,¹⁰ has studied with minute care a case of syringobulbia, that of a young girl who has been a patient of mine for some years at the National Hospital. In this case one of the earliest symptoms was a sensation of *intense coldness* over the right arm and leg and right half of the body, with similar coldness on the left side of the face. To the dysæsthesiæ there corresponded objectively loss of recognition of cold stimuli, which were either said to be neutral or misappreciated as warm. The loss to warm or hot stimuli was less in evidence; it was no more than partial, for impulses from very hot contacts were correctly recognized, though merely as "warm." In this respect, therefore, Jackson's hypothesis was to some extent confirmed.

Again, two of my former assistants, Drs. D. R. Gowler and B. M. Hope,¹¹ have published the case of a patient of mine at King's College Hospital, which is one of occlusion of the right posterior inferior cerebellar artery. In this instance the patient complained of an *intense burning pain* over the whole of the left side of the body and left limbs, and on both sides of the face, with severe "pins and needles" or painful tingling in the same distributions. Objectively, heat stimuli (about 45° C.) were recognized on both sides of the body and face, whereas cold (about 7° C.) were completely missed on the left half of the body (except on the neck), and left limbs. There was, however, no impairment of the appreciation of cold on the right side of the face. Thus (1) the burning dysæsthesia of the left body and limbs coexisted with objective loss of sensibility to cold in the same areas and with conservation of sensibility to heat (for Hughlings Jackson's view); but (2) it occurred over the face on both sides where no loss of either form of thermal experience could be found (against Hughlings Jackson's view).

A number of years ago I¹² reported an example of the same lesion in which loss of appreciation of all degrees of cold over the right side of the body, right limbs, and both sides of the face was associated with "hot pins and needles," with "tingling warmth" over the same side of the body and same limbs, yet only on the left half of the face, whereas all degrees of heat were stated (for the right limbs, body and face) to be "tepid" or "slightly warm," and on the left face they were unrecognized. This case, therefore, illustrates the same peculiar correspondence and non-correspondence, at one and

the same time, of subjective thermal sensation and objective thermal loss, as does that of Gowler and Hope.

But it is valuable from another point of view. I quote from the original description :

CASE 12.—“ Mr. M. had often noticed that *pricking of the left face*, in addition to its being felt as pressure merely, *never resulted in any bleeding*. He had frequently experimented with himself in this connection, and says the same thing is true of the right side of the body. About three weeks ago, however, coincident with the diminution of the ‘ subjective sensations ’ on the left face, he found that pricking was followed by bleeding for the first time since the onset of the disease. I re-examined him from this point of view, and can vouch for the fact that blood is much more readily obtained by pricking on the left hand than on the right, whereas there is no difference now between the two sides of the face. There has never been any appreciable difference between the surface temperatures of the two sides of the body.”

That some vasomotor change had occurred over the left face and right body and limbs seems absolutely certain—presumably a degree of general vasoconstriction. Further (to quote again) :

“ The return of a certain vasodilatation, as indicated by bleeding after pricks on the left face . . . coincides with a diminution of the ‘ subjective sensations ’ of tingling when the skin of the left face is rubbed, and suggests that there may be some connection between the two, especially as these sensations remain undiminished over the right limbs, and here pin-pricks are not followed by bleeding.”

I am not aware that other recorded examples of occlusion of the posterior inferior cerebellar artery, or of other organic lesions in medulla, pons, or mesencephalon, have been specially studied from this particular standpoint, but my case does undoubtedly provide material in support of what I have already suggested, that there is a definite relation in some cases between spontaneous dysæsthesiæ and vascular (vasomotor, neurosympathetic) disorder.

Finally, in respect of the brain-stem level, I have discovered a striking instance of a lesion of the posterior inferior cerebellar artery, recorded more than one hundred years ago by Alexander Marcet, M.D.,¹³ in the second volume of the *Medico-Chirurgical Transactions*, 1811, under the heading “ History of a singular nervous or paralytic affection, attended with anomalous morbid sensations.”

CASE 13 (Marcet).—The case is that of Dr. Vieusseux, age 62, who had an ictus on January 4, 1808. Thereafter, he was found to show complete analgesia of the right limbs and right side of the body up to the

neck, and the left half of the head was also analgesic. Further, thermal appreciation was gravely disordered, as can be gathered from the following excerpts which I have selected.

"An attendant brought him an etherized julep, which he took with his right hand, and the bottle felt lukewarm; but on taking hold of it with the left hand he found it cold, as it really was."

"On putting his right hand into boiling water it appeared so far from hot that he could have kept it immersed without being sensible of its scalding him, had not a disagreeable sensation, different from that of burning, at length warned him to withdraw it." "In plunging into very cold water, the water appeared almost warm to his right side, but very cold to the other."

"There was constantly a *sense of heat* throughout that (right) side, often as if by puffs, or as if hot cloths were suddenly applied to the parts."

"He often had a *sensation of cold water* all over his face, especially when in the open air, which induced him to wipe himself as if he had been wet."

I need not cite further from this century-old case the graphic details furnished by the self-observation of one of our own profession, and recorded by another with minute accuracy; their work lives after them and enables us three generations later to recognize the unmistakable symptoms and signs of a lesion of the left posterior inferior cerebellar artery. (1) We here meet with a thermal dysæsthesia of warmth or heat on the right side, with absolute loss of the opposite mode (cold) on the same side, and with loss also of appreciation of heat. Yet one should note that these two thermal losses were not identical in all respects: "On putting him into a cold bed, it appeared hot to the right side, and cold to the left. In getting into a hot bath, it felt hot to the left side, and neither hot nor cold to the right." That is to say, on the affected side cold things felt hot, but hot things did not feel cold. This distinction, existing with the subjective sensation of warmth or heat, is precisely identical with what obtains in my Case 9 (above), as the following remarks of my patient, given verbatim from the case record, show: "If I get into a cold bath the water would feel cold to the left leg and hot to the right; if I lie in a hot bath the water feels hot to my left leg, but I do not feel the water at all with my right leg." To the possible meaning of this interesting interrelation for the problem of the physiology of dysæsthesiæ reference is made at a later stage. (2) Marcet's patient also exhibited the phenomenon of subjective wetness on both sides of the face, but from the record it cannot seemingly be linked to any definite objective sensory change on both sides; apparently the right face was normal, while

on the left side, in addition to the analgesia already mentioned, there was "insensibility to ice." Touch, seemingly, was everywhere intact.

(4) THALAMIC LEVEL

"Subjective sensations" constitute an integral part of the familiar thalamic syndrome, the establishment of which we owe largely to the successive work of Dejerine,¹⁴ Roussy,¹⁵ and Head and Holmes.¹⁶ Whether they assume the form of spontaneous pains and painful paræsthesiæ, or of thermal or tactile or mixed dysæsthesiæ, they differ in no material respect from those met with at all the lower levels. Both the hyperalgesia and the hyperpathia which we have seen may result from lesions below the thalamus can similarly characterize affections of the latter ganglion. None the less, thalamic lesions of the class producing the thalamic syndrome proper can be recognized by the occurrence of clinical details which need not here be particularized.

In view of the confluence of the fillet sensory systems on the thalamus and their ending in its lower (ventrolateral) aspect, a degree of mixture of spontaneous sensations is naturally to be expected. If the unbearable pains of the typical syndrome usually predominate, drowning other sensory elements in consciousness by their sheer intensity, yet in one or other case some other variety may either coexist or actually be more pronounced. With these subjective combinations objective variations, ranging from hyperalgesia to analgesia and with corresponding extremes in other sensory divisions, can be observed. For present purposes one or two examples of the syndrome exemplifying other dysæsthesiæ than that of pain alone will be chosen.

CASE 14.—A. P., male, age 52. Diagnosis, lesion of right optic thalamus ; thalamic syndrome.

The patient had a stroke eighteen months ago, causing a mild left hemiparesis and a much more evident sensory hemiplegia. He complains of insufferable *pins and needles* all over the left side ; "I feel as if I were treading on tin-tacks." At one time there is a pricking, *burning* sensation in the left face, trunk and limbs ; at another it is *icy cold*. Objectively, a characteristic hyperpathic response of an explosive nature is obtained, more especially with pin stimuli ; cotton-wool touch is diminished ; cold excitations fail to reach the sensorium, while heat or warmth is much better appreciated. There is no astereognosis, loss of muscular sense, or defect of localization, but the phenomenon of radiation is pronounced. In respect of pain, the "all-or-none" nature of the response prevails ; a single mild pin-prick becomes a "stab," and

is felt "as if the pin were being dragged deeply along." If not a "stab," it is never felt at all.

This case illustrates painful, thermal and tactile dysæsthesiæ in one subject.

CASE 15.—A. R., female, age 53. Diagnosis, thalamic syndrome with athetoid movements.

Eighteen months before coming under observation at the National Hospital, the patient had a stroke (embolic) involving the right side and was unconscious for three days. Multiplicity and inconstancy of duration and degree have been the salient features of the dysæsthesiæ.

Thus :—

(1) *Numbness* and "deadness" of the right face, trunk, and limbs is often remarked.

(2) *Tingling* and *pricking* also, but not pronounced.

(3) More persistent is a *burning* feeling all down that side, "something cruel"; on one occasion it was so extreme she actually "thought the bed was on fire," and looked to see.

(4) Again, this is replaced by a *cold* sensation over the right face and limbs, especially the upper, but not the trunk: "just like *pouring cold water* down over the face, neck and arm"; hence a feeling of "rushing" or "moving" goes with it.

The right arm and hand are the seat of unceasing athetoid movements, and the plantar response is extensor.

Objectively, tactile and painful sensibilities are both reduced, thermal (both forms) also, but heat less than cold. Hyperpathia for painful stimuli is much in evidence, and "burning" usually goes with it.

Case 15 similarly exemplifies well the complexity and multiplicity of the subjective sensations associated with some organic lesions at the level of the optic thalamus. Other cases of the thalamic syndrome exhibit the phenomenon of *alloparalgia* already alluded to under the section dealing with *causalgia*. Some examination of the hypotheses advanced to account for the nature of the dysæsthesiæ associated with thalamic disease can be made more conveniently after our résumé of the data of all the sensory levels is completed.

(5) CORTICAL LEVEL

Leaving aside, therefore, for the moment the vexed question of whether the optic thalamus is a *terminus ad quem* for at least one form of sensory experience, viz., that of pain, we must ascertain whether with known cortical lesions *paræsthesiæ* or *dysæsthesiæ* can occur, and if so, of what qualities. We are here dealing, be it remembered, with the relatively crude, simple, separable varieties

of mere sensation with which an affective element of pleasantness or unpleasantness is inextricably combined, and not with those definitely higher, more complicated, perceptions which all believe are of cortical site, physiologically speaking, and which in reality partake of the nature of discriminative judgments, e.g., stereognosis and other sensory syntheses—in Parson's ¹ terminology, "highly apocritized perceptions." Is there satisfactory clinical evidence of the origination of dysæsthesiæ of tactile, painful, thermal, or mixed type from lesions in which only the cortex is involved?

Evidence of the occurrence of both subjective and objective alteration in sensation accompanying lesions of cortical site has accumulated over many years, and has been of late excellently recapitulated by Piéron ² and by Foerster, ³ to whose books the student of the subject is referred. The experimental evidence of Cushing, ¹⁷ derived from operation cases in which the cortex has been stimulated in the conscious state, and of others who have made similar investigations; the animal experiments of Dusser de Barenne ¹⁸ and of Minkowski ¹⁹; the material derived from cases of gunshot wounds in war; and, not least, that obtained by correlation of clinical symptoms with pathological findings (tumours, vascular disease), all combine to establish the fact that varieties of common sensation can arise from cortical excitation or irritation, and be lost from cortical destructive lesions. Of the genuineness of this conclusion the clinician has long been convinced. It appears that the representations in the cortex of varieties of sensibility are distinctly unequal, some being more perfect than others. Both of the above-mentioned authorities quote researches of their own, and numerous other published reports, which go to show that pain has a definite representation in the human cortex, as well as thermal sensibility, and that previous conclusions in the opposite sense must be reconsidered. Piéron, for instance, has worked out ingeniously in statistical form the frequency and intensity of injuries (in cortical war cases) to the simple forms of sensibility and elementary modes of perception. Attributing to each sensation a coefficient running from 1 (considered normal) to 5 (complete disappearance), he finds that heat and cold are least affected (3·21), and pain comes next (3·35). Tactile discrimination (3·80), position (4·00), and stereognosis (4·13) are most affected.

The clinician can furnish collateral evidence, as far as the dysæsthesiæ are concerned, derived from the study of cases of Jacksonian epilepsy. Cases of this category in which paræsthesiæ

in a limb or limb-segment usher in the epileptiform attack, commonly of the nature of tingling, or of something "drawing" or "moving," are very frequent. I have not, to the best of my recollection, noted a case of sensory aura amounting to *pain*, but tingling or "pins and needles" is not far removed therefrom, while Foerster has observed a case of Jacksonian epilepsy the invariable aura of which was pains in bladder and rectum, followed by paræsthesiæ on one side, and he allows definitely the occurrence of such subjective pain of cortical origin. The following case, also of Jacksonian epilepsy, illustrates the definite development of *thermal* dysæsthesiæ as a cortical aura.

CASE 16.—V. F., female, age 39. Diagnosis: right parietal cerebral tumour (successfully removed).

This patient suffered from repeated Jacksonian attacks involving the left limbs, beginning in the left hand and spreading quickly to the whole of the left side. *The aura was a subjective sensation of icy coldness down the left side (face, trunk, limbs), which appeared suddenly a minute or two before the motor symptoms, and remained till they developed, passing off with their appearance.* In addition, frequent pains were complained of in the left limbs (not as an aura, but apart from the attack). Objectively, the left hand exhibited astereognosis and loss of muscular sense; touch was lost over the major part of the left side, and pin-prick stimuli were diminished (not lost) distally in arm and leg.

Because of its historical and intrinsic interest reference may at this point be made to the case of Sir Philip Broke, quoted by Weir Mitchell ²⁰ under the heading, "Subjective false sensations of cold."

CASE 17 (from Weir Mitchell).—Sir Philip Broke fell from his horse on August 8, 1820, was concussed, if not completely unconscious, and suffered from confusion, amnesia, and vomiting. "The first symptoms that I remember of any affection of the nerves were my perceiving, in the afternoon of the day following the accident, a sense of *extreme cold* in my leg and foot and left hand, so that I could not sleep in comfort without a worsted glove and worsted stocking; and in the course of the next day I discovered that the whole of the left side was strangely affected, the sense of cold appearing to lie internally upon the coating of the bones of the arm, thigh, and leg; and that, though the flesh externally was warm to the touch and generally in a state of perspiration, and though the skin appeared perfectly fresh and smooth, yet . . . the whole left side of my person was affected with a singular numbness to the touch."

Sir Philip was seen subsequently by Sir Astley Cooper, who diagnosed "an extravasation of blood upon the brain or its membranes." Further details are given in Guthrie's *Military Surgery*.

"The concussion of the brain . . . rendered the left half of his whole person incapable of resisting cold or of evolving heat. In a still atmos-

phere abroad, at 68° F., he said, 'the left side requires four coatings of stout flannel, which are augmented as the thermometer descends every two degrees and a half, to prevent a painful sense of cold, so that when it stands at freezing-point the quantity of clothing of the affected side becomes extremely burdensome.' "

Paucity of detail renders the century-old account less informative than is desirable ; yet we may legitimately infer that the symptoms were attributable to a hæmorrhagic effusion, which is at least as likely to have been cortical or submeningeal as intracerebral ; hence the case provides another example of thermal dysæsthesiæ of cerebral and possibly actually of cortical origin.

No case of *wetness* as a cortically arising dysæsthesia has as yet come under my notice, and I shall not now refer to more elaborate forms of subjective sensation, interesting though they are, constituted by hallucinations of the muscular sense—in which, for example, the Jacksonian sensory aura takes the form of a sensation of the arm moving or rising into the air, while in actual fact it does not thus move at all. More than one case of this rare kind I have had occasion to observe.

We have now concluded our sketch of the spontaneous or subjective sensations making their appearance in association with organic disease at all physiological levels, from peripheral to cortical, and the task that remains is to attempt a synthesis of their pathogeny and an investigation of their neural correlates, as far as may be practicable in the present state of knowledge.

Pathogenesis

Since the dysæsthesiæ with which we have been dealing are all conscious sensations, it follows without question in each and every instance that afferent paths from the site of the lesion to the "perceiving cortex" must be open, and that the latter itself, wherever perception originates therein, is in a state of integrity. If *cortical* disorder of function causes a dysæsthesia to arise in consciousness, the route from its particular site to the "perceiving cortex," and the latter, must be not disordered.

EXCITATION AND INHIBITION

The pathogenic problem of irritation versus removal of inhibition or release of function seems unavoidably, perhaps, to arise at the outset—though whether these apparently opposite processes should

be considered radically different is not at present altogether clear to me. It is incontrovertible that stimulation can and does awaken dysæsthesiæ in consciousness. Direct excitation of peripheral nerve-trunks (mechanical, chemical, electrical, etc.) causes pain, or, if weak, pain-like paræsthesiæ. Pressure, or a knock, on the ulnar nerve at the elbow invariably arouses painful tingling, and no other specific sensation. Noxæ of an organic kind seem also to act definitely as irritants—witness the pains of any toxic or toxoinfective agent affecting peripheral nerves. At a higher level root pains must in many instances be of purely excitatory origin, if we believe that compression irritates. Local injections of strychnine into the dorsal horns of the cord give rise to severe pain in corresponding segmental cutaneous zones (Dusser de Barenne ²¹), while, according to Foerster, electrical stimulation of the anterolateral funiculi results in pain, and of the columns of Goll in the cervical cord, in paræsthesiæ. Direct electrical excitation of the sensory cortex in conscious human subjects has often been shown to be accompanied by dysæsthesiæ, as already alluded to. Thus at all levels direct irritation of afferent systems must be allowed to be definitely algogenic or paræsthesigenic.

Again, the significance of the awakening or aggravation of dysæsthesiæ by extrinsic stimulation ought not to be underrated. Numerous instances of this have been met with by every clinician, and some have been narrated in the case-histories given above. If in a case of causalgia momentarily latent the application of some dry or warm substance arouses agonizing pain, it can be solely by way of excitation, at least to all appearance.

On the other hand, the clinician tends to become dissatisfied, rightly or wrongly, with hypotheses predicating the existence of persistent or permanent irritation, and, influenced by present-day neurophysiological conceptions, may seek rather to account for long-continued pains and dysæsthesiæ by some view explaining them as release-phenomena. To Sir Henry Head ²² we are indebted for an ingenious theory elucidating the central pains of certain thalamic lesions as release-phenomena rendered possible by removal of corticothalamic inhibition. We shall return to this matter shortly, but for the moment we must note the limitation of the conception to the pain-system and to thalamic cases. Whether it be not also of wider applicability is a point for discussion. Certainly I see no good reason why it, if accepted at all, should not also be utilized to account for persistent thermal and other dysæsthesiæ of central

origin. That thalamic activity should be accompanied by consciousness for one variety of sensory experience, but not for others, is a difficulty for Head's theory not lightly got over, "essential organ of the thalamus" notwithstanding.

For the sake of clarity let us take a concrete instance of persistent thermal dysæsthesia. Only a short time since I saw once more the patient with syringobulbia (case reported by Wyllie¹⁰) referred to above, and ascertained that the dysæsthesia of icy coldness on the right side remains as before; it has now continued for at least six years. Of the existence of a syringobulbic lesion involving the formatio reticularis of the left medulla there can be no question. Does the dysæsthesia persist because afferent fibres concerned with conduction of impulses underlying the sensation of cold (fibres anatomically interrupted by the lesion, since objective thermanæsthesia is present on the right side) are *in part* being "irritated" thereby, and the excitations interpreted in consciousness as a sensation of cold, with local signature? Or, alternatively, because a corticifugal inhibitory path, extending to the thalamus and beyond, is somehow impaired in function, allowing "hyperphysiological" afferents to pass to the thalamus and cortex? As for the latter conjecture, that of an efferent or corticifugal inhibitory system, it must extend lower than the thalamus if it is to be involved in the medullary lesion. What evidence is there of such descending inhibitory sensory systems, or what valid reason for the necessity or desirability of such a hypothesis? "Irritation" of the afferent sensory system concerned by the existing structural bulbar lesion surely must appear an at least equally feasible explanation, if not much more so. However, the problem can be approached from a somewhat different viewpoint.

DUAL MECHANISMS IN THE SENSORY SYSTEM

We owe to the profound researches of Head²² the idea of a dual mechanism in the sensory system taken as a whole, and while his conclusions have not received general acceptance, it is curiously interesting that a number of recent workers seem to adopt some kind of dualism, though not in accord with the original subdivisions of the former, and with no unanimity among themselves. Thus Parsons,¹ who (not entirely on the lines of Head) finds a fundamental contrast between what he calls "crude dyscritic sensibility" and "highly differentiated epicritic sensibility," declares that many facts "point to a dual mechanism at work in almost every domain

of sensation, *with the exception of pain* " (italics mine), and is led to the conclusion that "Head's distinction of protopathic and epicritic systems is fundamentally correct in spite of grave difficulties and inconsistencies in detail." There is no trace here, however, or in Head's scheme, as far as I can find specifically, of any conception that in a Jacksonian sense the two form a pair of corresponding opposites, and that one antagonizes the other—in other words, inhibits it. Foerster's latest contribution, however, to which I have several times already referred, embodies a theory of the following nature.

There is evidence, Foerster thinks, for the existence of two sensory systems: (1) *affective*, for the appreciation of pain and of pleasant and unpleasant qualities in sensation; (2) *perceptoric-epicritic*, for "pure sensation," viz., touch, pressure, warmth, cold, etc., unaccompanied by affective elements, and specially concerned with discrimination and spatial appreciation. Further, his speculation is to the effect that the former is normally regulated and inhibited by the latter; he concludes "dass normaliter das Schmerzsystem durch die gleichzeitige Erregung der Empfindungssysteme einer Regulation unterliegt, dass es einerseits eine Inhibition durch die Empfindungssysteme, andererseits eine Verfeinerung seiner Leistungen durch letztere erfährt." Consonant with this speculation, Foerster supposes one of the explanations of hyperpathia to reside in an overaction, a setting free, of the "pain-system," by impairment of the control normally exercised over it by the "sensation-system." This is an ingenious hypothesis, though it obviously cannot cover all the clinical phenomena of hyperpathia, nor is it by any means universally applicable to the conditions we are considering, in which, for example, one part or subdivision of Foerster's "sensation-system" *itself* is overfunctioning, or in which *both* systems are so in overaction (cf. the *burning* + the *pain* of *causalgia*).

But this is not his only idea of sensory inhibition. For spinal cases of spontaneous or central pains, he imagines a corticifugal path extending to the limiting layer in the lateral column, close to the dorsal horns, and normally exercising an inhibitory effect on the pain-system cells of these horns. Similarly, he imagines the existence of a corticifugal inhibiting system extending to the brain-stem, which is out of action in cases of central pain at bulbopontine levels; finally he alludes to the Head and Holmes' view of a cortico-thalamic system of inhibition, accepting it as a possibility, but not

declaring for it uniquely in explanation of thalamic pains. He considers, indeed, that thalamic activities are inhibited also from the corpus striatum.

Plausible as all this conjectural mechanism may possibly appear, after much consideration of the problems presented by the dysæsthesiæ I cannot see that it is either necessary or as explanatory as at first it may seem. I do not find it essential to posit an elaborate inhibitory system that passes out of action when painful or other varieties of dysæsthesia come into being. Qua pain, that of trigeminal neuralgia in no material way differs from central pain, and no one evidently is inclined to attribute it to removal by disease of an efferent pain-inhibiting system extending to the Gasserian ganglion; it can be explained more comfortably on the view that slow fibrous or other structural or toxi-infective changes in the ganglion irritate and excite the protoneurones. Where dysæsthesiæ are in evidence and objective sensory alterations cannot be detected, irritation of the sensory system and paths concerned seems to me feasible from the viewpoint of pathogenesis. If objective changes accompany the subjective phenomena, then the system involved is partly irritated and partly under destruction. If a form of anæsthesia dolorosa occurs (we might well use some similar expression for its analogues, such as subjective coldness with loss of appreciation of cold stimuli), we account for the loss by the block from the lesion, and for the dysæsthesia by "irritation," it may be, of fibres still capable of conducting excitations, which are misinterpreted by the cortex since they do not, as a fact, originate in cold applications. Obviously, at peripheral and at central levels, certain differences in the physiological process are to be expected.

CRITICISM OF A JACKSONIAN THEORY

The case is somewhat different, on the other hand, in respect of those examples (a few are given in this series) of subjective sensations of heat, or of cold, with objective loss of the *opposite* modality. Take concretely Case 7, in which subjective coldness was associated only with loss of heat appreciation. For cases of this category Hughlings Jackson⁸ offered the following theory: "The super-positive element of the double symptomatology, the over-coldness, is not caused by any disease of the physical bases of the sensation cold; obviously it cannot be if the disease has produced, as we are supposing, a negative lesion. The continual over-coldness is consequent on removal of control, synonymously of inhibition, from

the physical bases of the sensation cold, by destruction or by some negative lesion of the physical bases of the sensation heat ; there is a cessation of antagonism such that the nervous arrangements which are the physical bases of the sensation cold are ' let go,' and rise in activity . . . with an associated rise in the sensation cold, a continual over-coldness referred to the skin."

It should be remarked in this connexion that Jackson's view is stated by him for cerebral cases only, and that he does not apply it specifically to cases with organic lesions at lower levels, though there appears to be no objection against its thus being applied. The hypothesis presupposes an innate or inherent antagonism or mutual inhibition, as between the two modes of sensation, hot and cold, being corresponding opposites. As a part of a wider principle of physiological antagonism, the particular illustration given by Jackson bears closely on one aspect of the problems of dysæsthesia, yet its application must be allowed to be distinctly limited. As we have seen, clinical cases which do *not* substantiate it are no rarity. Again, the reader may recall that in one and the same case *both* correspondence *and* non-correspondence between dysæsthesia and loss of objective opposite can occur at the same time (cf. Case 12, and that of Gowler and Hope ¹¹). Finally, Jackson's hypothesis does not explain satisfactorily the objective loss of *both* modes of thermal sensibility in cutaneous areas where the patient has a subjective sensation of one only. Thus the patients in Case 9 and Case 13 felt neither the coldness of things cold nor the hotness of things hot over the region of the dysæsthesia of warmth or heat. While cold applications gave rise to a sensation of heat, hot applications felt neither hot (as they should have done according to the theory) nor cold. I submit a possible explanation is that the existing subjective hot sensation in consciousness prevented the subject from appreciating additional hotness effected by the application of hot substances to the skin areas concerned, on the one hand, while the loss of cold conductivity precluded the chance of their giving rise to a false sensation of cold on the other. We should also note the seeming failure of the hypothesis to explain the fact that, objectively, cold substances occasioned a sensation of warmth ; that they did not result in a cold sensation follows from the destruction of the " physical bases " of that sensation, and one may presume their wrong appreciation is somehow connected with the existing subjective " warm " or " hot " dysæsthesia.

Thus so complex are the clinical possibilities that no unitary

explanation covers all instances, apart from the consideration that physiological antagonisms in respect of other forms of sensory experience than the thermal do not suggest themselves with the same facility.

CORTICOTHALAMIC INHIBITION

While, then, a pathogenesis by stimulation or excitation seems to present fewer difficulties and be less hypothetical than that depending on the existence of inhibitory systems, the possibility of the latter being a well-founded explanation for some of the phenomena is not to be lightly dismissed, and may yet find ampler evidential support than it would appear to have to-day. The theory according to which the spontaneous pains of the thalamic syndrome are caused by removal of inhibitory influence of cortex on thalamus is perhaps the least unattractive of the inhibition hypotheses; but I do not wish to leave this part of my subject without some allusion to an obstacle in the way of its acceptance. At the beginning I made the elementary remark that we can only know of our patients' dysæsthesiæ by their verbal description of them. This fact has been made use of by Graham Brown²³ in a criticism of the theory of corticothalamic sensory inhibition, and I cannot do better than quote his exposition. The theory implies that a psychical function (that of the appreciation of pain) is localized in a subcortical region, and that thalamic activity (at least under some conditions) is accompanied by consciousness thereof.

"We may leave aside the assumptions that the descending influence of the cortex upon the thalamus is one of depression, and of depression only, and that sensibility to pain is not affected by a lesion of the cortex, however large it be. There remains the fundamental obstacle that the patient in question was able to overact by *speech*. If the thalamus itself takes part in the mechanism of the overaction, *it must therefore still be in connection with the speech mechanism*. This fact forces us to one of two conclusions. Either the motor speech mechanism lies in the thalamus, and the thalamus in that case itself 'said' that it overacted; or that mechanism lies in the cerebral cortex (or in some other region of the cerebrum than the thalamus), in which case the 'essential organ' of the thalamus turns out to be merely a portion of, or a relay station in, the path between receptor and motor speech mechanism. In this latter case the site of the overaction *may* be in the thalamus (if the lesion removes an inhibitory influence of cortex upon *thalamus*); or it *may* be in the cerebral cortex (if the inhibitory influence removed is one of thalamus upon *cortex*). . . . Even if we were able accurately to localize the site of the overaction, we should still be as far as ever from localization of the 'site of consciousness' for affective sensations. We may just as

legitimately suppose it to be in the speech mechanism (or other motor mechanism) as in any preceding path of conduction" (*italics in original*).

We need not here enter on questions of awareness and degrees of consciousness, nor need we, at the same time, hesitate to entertain the possibility of psychical phenomena (of a graded kind) accompanying physiological activity at lower levels than the cortex and in simpler organisms than man, but as far as cutting off of thalamus from cortical control is concerned, it *is* connected with the cortex if the patient reacts by speech, and therefore *consciousness* of the sensations of pain *may* have another site than the thalamic "essential organ."

THE NEUROVASCULAR FACTOR IN PATHOGENESIS

From various remarks already made apropos of a number of cases in this series, the reader will have gathered that another pathogenic factor than excitation or inhibition of neurones belonging to the nervous system proper may be responsible for some at least of the dysæsthesiæ. The association of one or other type with lesions (presumed or proved) either of sympathetic nervous system or of vascular system or both, is manifested in Cases 4, 5, 9, and 12, and may be justifiably surmised in others. I am myself convinced of the etiological significance of the relationship, which is strengthened by knowledge of a number of other clinical illustrations (personal and communicated). By kind permission of Professor E. Archibald, of McGill University, Montreal, I am allowed to refer to an unpublished case of his, that of an elderly gentleman with persistent pain in the stump of an amputated thigh, for which all the usual medical and surgical procedures had proved unavailing until Professor Archibald performed a periarterial sympathectomy on the femoral artery in Scarpa's triangle, when the pain ceased and has never returned.

The lines of evidence may be marshalled somewhat as follows.

(1) Blood-vessels normally are sensitive to pain. In Foerster's experience ligation of carotid, external iliac, vertebral, brachial and popliteal arteries is accompanied by pain. Fine neural networks have been demonstrated in the adventitia, and sensory end-plates on the external walls of arteries. The phenomena of migraine were shown many years ago by Lauder Brunton ²⁴ to have a basis in angiospasm or vasoconstriction, while conversely migrainous pain ceases with the injection of angiodilators.

(2) Sensory nerves of vessels run in periarterial networks, passing

therefrom at intervals to join main nerve-trunks (Kramer and Todd,²⁵ Potts²⁶). The possibility of a continuous side-path for vessel pain, constituted by afferent fibres which do not unite with nerve-trunks but pass directly to the sympathetic paravertebral chain, eventually to enter the neuraxis at higher levels, is denied by most but accepted by Foerster, who relies to some extent on the following experience of his own.

The case was one of complete transverse lesion of the three lower roots of the brachial plexus (C 7, C 8, D 1), with complete loss of all forms of cutaneous sensibility over the three ulnar fingers and the ulnar side of the hand. Deep pain, however, was preserved. Foerster exposed the nerv. digit. vol. proprius on one side of the little finger and stimulated it with a powerful faradic current, without the slightest consciousness of pain on the patient's part; but when he applied the electrodes to the corresponding digital artery the pain was intense.

It would appear, therefore, that three afferent pain paths may exist: (a) peripheral nerves, posterior spinal roots, cord; (b) vessel plexuses, peripheral nerves, spinal roots; (c) vessel plexuses, aorta, sympathetic paravertebral chain, white rami, posterior roots.

(3) The evidence furnished by cases of periarterial sympathectomy (Leriche,²⁷ Müller,²⁸ Platon,²⁹ Quarry Wood,³⁰ and many more) points undoubtedly to the modification of certain sensory disorders by operation on arterial plexuses, apparently by removal of angiospasm and development of relative hyperæmia. This clinical evidence must, in my opinion, be accepted, however obscure the exact mechanism of the production of the phenomena may still be. Indeed, an anatomist like Ranson³¹ allows himself to say "it is impossible to explain the relief from pain which some patients experience after this operation" (periarterial sympathectomy). In such instances, as in others, clinical research has preceded that of anatomy and physiology.

(4) The pronounced dysæsthesiæ of Case 12 were present over a wide cutaneous region in which a concomitant angiospasm of central origin existed, so that pin-pricking did not result in bleeding. But with diminution in intensity of the subjective sensations return of bleeding on pin-prick took place. This association was much too definite to be merely coincidental, apart from the fact that it is borne out by cases of hysteria, as the following case, taken from a former article of mine,³² clearly proves.

The patient was a young woman with hysterical paraplegia, and with numbness of the legs corresponding. So absolute was the analgesia objec-

tively that a needle was passed through the calf without its being perceived, and without causing any bleeding.

"After a strong application of the wire brush the analgesia entirely disappeared, and where a minute or two before she had felt nothing she now felt the needle instantaneously; and, in addition, *where formerly a vasomotor defect had prevented bleeding there was now bleeding whenever the skin was pricked.*"

These considerations, together with others which must be here omitted, unquestionably point to the development of dysæsthesiæ (of some kinds) as a sequel to vascular disorder of sympathetic origin. The clinician does not pay sufficient attention to the occurrence of venous neuralgia (cf. Edinger³³), or of paræsthesiæ plausibly ascribable to changes in the vasa nervorum (cf. acroparæsthesia, above). So far as precise knowledge takes us, on the other hand, it seems rather premature to assign the reception of pain *uniquely* to sympathetic nerve-endings in skin and muscle, as some are inclined to do. At the same time, we should not underrate the possible significance of the presence of unmyelinated nerve-fibrils as well as of myelinated fibres in most sensory end-organs (Agduhr and many others). Further, the histological researches of Boeke³⁴ appear to prove the existence of a sort of cell-syncytium in the end-organ, into which the periterminal network of fibrils passes by stages so gradual that it is sometimes difficult to say where fibril ends and cytoplasm begins. Thus the possibility of sensory end-organs being influenced peripherally by local blood-states is more feasible than a theory such as that of Trotter³⁵ would allow; according to the latter the nervous system is insulated from mesodermal (inter alia, vascular) tissues by various forms of laminated capsule (for the nerve-endings), and by perivascular sheaths (for the neuraxis). Histological discoveries rather militate against this view, as we have seen above at more than one stage in our discussion.

Summarizing, we may, I think, conclude that under certain circumstances a neural origin, under others a vascular or sympathico-vascular origin, underlies the manifestation in consciousness of tactile, painful, and thermal dysæsthesiæ.

EXPLANATION OF HYGRIC DYSÆSTHESIÆ

The interpretation of dysæsthesiæ of a mixed type, such as that of wetness, is more complex. Cases 2, 11, and 13 exemplify its genesis at peripheral and bulbar levels—so far its appearance at higher levels has not come under my notice. References to the

phenomenon in the literature are peculiarly difficult to find. Parkes Weber ³⁶ has recorded a case of splenomegalic polycythæmia in which the patient said that "whatever she touches with her hands (when her hands are cold, as they often are) feels wet to her." I have discovered a number of allusions to this "hygric illusion" or paræsthesia (sensation de mouillure) in older French and Italian literature (Ramadier,³⁷ Mingazzini,³⁸ Giannuli,³⁹ Peli,⁴⁰ Ravenna and Montagnini ⁴¹). The majority of the reported cases concerned patients who were definitely alienated, and the phenomena were rather of the class of hallucination and presumably of psychogenic derivation. Attempts were made by more than one of these authors to localize the "hygric illusions" in the gyrus hippocampi, but the pathological evidence offered is unimpressive.

Introspectively, one may consider a hygric dysæsthesia compound of a tactile and a thermal element, to which an element as of "something moving" may perhaps be added. Immersing one's forearm in water one is conscious of: (1) a tactile ring at the level of its surface; (2) a sensation of warmth or of coolness; (3) a general sensation of touch-pressure over the whole cutaneous area immersed. When a patient describes the sensation as consisting also in a feeling of "something trickling" the element of "movement" certainly contributes to the whole complex. On the other hand, when (as in Cases 2 and 13) the complaint is of a feeling of wetness on the face I do not see that a thermal component is necessarily superadded, for objective perspiration can arouse this wet feeling, and in these circumstances it would seem that there must be patchy excitation of touch-spots over areas of measurable extent, and again, possibly, the above-mentioned feeling of "movement" on or over the skin. Relative juxtaposition at peripheral or bulbar levels of touch and thermæsthesia-conducting tracts may explain the origin of this complex dysæsthesia; at the level of the cortex their separation possibly prevents its occurrence as of "central origin"; and, theoretically at least, it should also on occasion be thalamic, but I have found no reference to such, nor as yet encountered any such case personally.

As remarked at the outset, my purpose has been to deal mainly or solely with dysæsthesiæ in association with organic or structural conditions, and in consequence nothing has been said of the "neurasthenic" and "functional" varieties of "pressure," "crawling," "drawing," "dragging," "tightness" ("as if in a vice"), etc., or of even more elaborate nature. Not that all or any of these are

specifically "functional," for such is not the case, but one must set limits to the present discussion of a particularly wide-ranging and complicated topic. I have long been convinced of the desirability of observing and analysing the *apparently* spontaneous dysæsthesiæ described by neurotic or psychotic subjects, and commend to the reader the philosophic interpretation of these undertaken a number of years ago by Dana.⁴²

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CHAPTER XIV

THE ARGYLL ROBERTSON PUPIL *

Definition. Clinical Conditions in which the A.R. Sign is found: Neurosyphilis; Epidemic Encephalitis; Disseminated Sclerosis; Mesencephalic Tumours; Traumatic Cases. The Anatomo-physiological Arc for the Light Reflex. The Path for Convergence and Accommodation. Site of the Lesion underlying the A.R. Sign. The Myosis of some A.R. Pupils.

Contributions to the study of the Argyll Robertson phenomenon have been so frequently introduced by allusions to its prominent rank as one of the vexed questions of neurology that omission of such preliminary matter may be not unwelcome. If, none the less, the problem still awaits full solution, the lines of approach have been laid down, and clinicopathological as well as experimental data, whose import is not to be mistaken, have been steadily accumulating. In this chapter fresh evidence is furnished pointing to the central origin of the disorder, and a new and simple explanation is proffered for its common occurrence in neurosyphilis.

Definition

Strangely enough, doubt still exists as to what actually constitutes the Argyll Robertson pupil, and unless unanimity is reached such horrid expressions as "a pseudo-Argyll" will continue to blot the pages of what is euphemistically called medical literature. The point is simply whether myosis is or is not to be taken as an integral part of the symptom-complex. Under the title "Four cases of spinal myosis, with remarks on the action of light on the pupil," Argyll Robertson ¹ published in 1869 a series of cases, chiefly of tabes dorsalis, in which he observed the phenomenon since associated with his name. Present-day opinion, however, with few exceptions, holds myosis to be incidental, so that the sign may be defined as consisting in absence (or obvious diminution) of the direct reflex to light, the

* *Journal of Neurology and Psychopathology*, May, 1921, vol. ii, p. 1.

consensual reflex being either absent or present, with preservation of the pupillary reaction on convergence-accommodation. That this is the soundest view to take is substantiated by the fact that myosis may occur without the dissociated reflex * phenomenon of the A.R. pupil, and vice versa, and since two different mechanisms are involved they should be considered separately. Why some 30 per cent. or more of A.R. pupils should also be myotic is a point of legitimate importance, discussed below. Vision is assumed more or less tacitly to be unimpaired in the A.R. phenomenon, but there seems no good reason to exclude cases of relative blindness which exhibit the dissociated reflex, though such are here omitted. A frequent but not perhaps constant correlated feature of the A.R. phenomenon is absence of dilatation of the affected pupil on painful stimuli from the trigeminal area or elsewhere. Irregularity or inequality of the pupils in the condition is incidental.

Clinical Conditions in which the A.R. Sign is found

1. The A.R. pupil thus defined, our first concern is with its clinical incidence. As every one knows, the A.R. phenomenon is encountered in a high percentage of cases of neurosyphilis, of whatever variety, though it cannot be taken as pathognomonic of that morbid condition, or as an infallible index to preceding syphilis. Recall of the prime neurological principle that symptoms depend less on the nature of the pathological process in the nervous system than on its site and the mechanisms involved should have led to hesitation in assuming a unipathological basis for the sign. As neurosyphilis is essentially a diffuse toxi-infective state, the peculiarly specific and local action of the spirochæte or its toxin on a particular mechanism will require an explanation which is both simple (for the A.R. pupil is very common in neurosyphilis) and in harmony with the occurrence of the sign in non-syphilitic cases.

By way of illustration only a single example need be given, selected because of the combination of unilateral myosis and bilateral A.R. pupil.

CASE 1.—S. S., female, age 40, has a strongly positive Wassermann reaction in the blood, but no signs in the nervous system except a double

* I use the expression "dissociated reflex" only for convenience of description; the reaction of the pupil on accommodation-convergence is not a "reflex" in the ordinary sense, but rather a part of a synkinesis, an "associated movement."

A.R. phenomenon. The right pupil is in a state of myosis and measures $1\frac{1}{2}$ mm. in diameter ; the left, on the contrary, is rather large, in diameter 5 mm. ; yet both show the typical dissociated reflex, and from neither is a consensual light reaction obtained in the other.

This case illustrates the impracticability of attempting to restrict the A.R. sign to the myotic eye, for in all other respects the phenomenon is identical in the two.

2. Of no less importance, if of minor frequency, is the occurrence of the sign in cases of nervous disease independent of syphilis. Still confining our attention to the toxi-infective group, we must emphasize its occasional appearance in the course of epidemic encephalitis. Tilney and Howe² state that manifestations due to interference with the oculomotor apparatus have been present in 53 per cent. of reported cases, and specify the occurrence of unilateral or bilateral paralysis of pupil-movement on accommodation, dissociated from light-reflex involvement—i.e., the reverse of the A.R. phenomenon, in a sense, for they do not point out that the former may be merely the sequel to a paralysis of convergence. The true reversed A.R. pupil consists in absence of reaction on actual convergence of the globes, with presence of the pupillomotor reflex to light. I have, however, seen at least one case of A.R. pupil in the disease.

CASE 2.—In a paper³ on epidemic encephalitis a case was detailed, of the mild recovering type, in which bilateral ophthalmoplegia externa was an early and marked symptom. The pupils, rather large and slightly unequal, reacted to light very sluggishly indeed ; convergence was impossible, nor was there any pupil movement in the attempt. At a later stage, however, uninterrupted improvement resulted in the complete disappearance of the external ophthalmoplegia, but the light reaction remained greatly impaired. After the paper was written I saw the patient on several occasions, and was able to satisfy myself that the response on convergence had become normal while the reflex to light was still very imperfect. That is to say, the condition approximated closely to the ordinary A.R. phenomenon. Both in blood and spinal fluid syphilitic tests always proved negative.

The A.R. pupil has since been recorded in the disease by Worms, Economo, G. W. Hall,⁴ Krabbe,⁵ and others.

It is known, of course, that reaction of the pupil may take place in an eye paralysed for convergence, on an attempt at the latter being made. Thus Guillain⁶ has reported a case of unilateral A.R. sign in a patient with a typical syndrome of Weber (non-syphilitic) ; the left eye showed the A.R. phenomenon, and was also the one

paralysed by the oculomotor lesion. When the patient converged, the right eye moved in and its pupil contracted, and simultaneously the left pupil contracted strongly, though that eyeball did not move at all.

3. Again, the sign has been observed occasionally in unmistakable cases of disseminated sclerosis. It was found by Uhthoff⁷ only once in one hundred cases, but it has been seen also by Probst,⁸ Pini,⁹ Marburg,¹⁰ Rad,¹¹ Liwschütz,¹² and others. Rad's two cases, in particular, do not appear to leave any room for doubt. I have myself seen one typical example in the disease.

CASE 3.—The patient was a young girl of 22, who came to the National Hospital complaining of paræsthesiæ in the limbs and of dimness of vision of subacute onset. On examination, the abdominal reflexes were absent and the plantars were in extension. Nystagmoid jerking was noted on lateral deviation. Investigation of the blood for syphilis proved negative.

Both pupils were rather dilated (diameter 5 mm.) and completely inactive to strong light, direct or consensual, but they reacted quickly on convergence. The optic discs were normal. I showed the patient to my colleague, Dr. James Collier, who corroborated the A.R. finding, and agreed that the case was one of early disseminated sclerosis.

From the foregoing it is apparent that a characteristic A.R. phenomenon is an occasional occurrence in diffuse toxi-infective states other than neurosyphilis, and surprise might be justifiably expressed if this were not so, for diffuse morbid processes must sometimes light on the same area and disturb the same mechanisms as are implicated in the A.R. pupil of syphilis.

4. The not infrequent development of the dissociated reflex, as a stage towards, or without going as far as, complete fixity of the pupil, has long been recognized to accompany certain cases of cerebral tumour in the vicinity of the third ventricle, aqueduct, or anterior corpora quadrigemina; but this knowledge does not appear to be widely diffused outside neurological circles. Yet these are the cases which furnish the most valuable clue to the general, i.e., commonest, site of the lesion underlying the A.R. sign, and are of significance out of all proportion to their comparative rarity. Two or three illustrative instances from personal observation may be given in some detail.

CASE 4.—A. B., male, age 19, came to hospital complaining of headache, lassitude, inclination to sleep, vomiting, diplopia, and uncertainty of gait. On examination, intense double papillœdema was found (highest

point + 6 D); there was no ptosis or nystagmus, but conjugate upward movement was defective, and there appeared to be some slight general weakness of the right limbs; cutaneous and deep reflexes alike, however, showed no deviation from the normal.

The pupils were of medium size; the left was inactive to light, and the right responded with great sluggishness, while both reacted briskly on convergence, i.e., there was a double A.R. pupil.

From the defect of upward movement the lesion was with some certainty attributed to the region of the anterior corpora quadrigemina, and it was held that the double A.R. pupil supported this localization. Improving somewhat under general treatment, the patient left without operation after a stay of some ten weeks.

He was seen again almost exactly four years later, and reported himself as quite well and at work. The optic discs were normal, with normal physiological pits. Conjugate upward movement was greatly impaired, in fact almost lost, the eyes tending instead to converge with the effort. No other symptom was present, except that both pupils were slightly but definitely irregular, medium in size, and exhibited a typical dissociated A.R. reaction on both sides, being immobile to bright light, direct and consensual, but responding actively with accommodation-convergence. On this occasion opportunity was taken to examine the blood for the Wassermann reaction, which was found to be negative.

No doubt can be entertained, I consider, that the case was one of cerebral tumour in the neighbourhood of the superior colliculi, the paralysis of conjugate upward movement being highly characteristic of lesions involving that region, and the accompanying typical A.R. sign must similarly be attributed to involvement of some structure or structures in the same part of the mesencephalon.

CASE 5.—C. D., male, age 42, was admitted to hospital with a six months' history of headache, giddiness, diplopia, staggering gait and a tendency to deviate to the left, and increasing drowsiness and apathy. On examination the patient was found to have advancing papilloedema in both eyes, highest point + 5 D. There was gross defect of upward movement, the effort always leading to slight divergence merely, with closure of the eyes. Convergence was fair on both sides, though less on the left than on the right. The pupils were equal in size, diameter $2\frac{1}{2}$ mm., and immobile to light, consensual or direct; but both reacted through a fair range, though not briskly, on convergence. No dilatation of either pupil was obtained on painful stimulation of the skin of neck or cheek. A decompression operation was undertaken without any resultant improvement, and some weeks later death ensued.

At autopsy a tumour was found involving both anterior corpora quadrigemina and the vicinity of the aqueduct generally, which proved on microscopical examination to be a glioma.

In this case the same combination of double A.R. pupil and

paralysis of upward conjugate movement of the eyeballs occurred, and the diagnosis of tumour of the superior colliculi was amply confirmed by subsequent pathological investigation.

CASE 6.—E. F., male, age 24, was admitted to hospital with a history that for three months he had suffered from headache, and that during the last week or two his vision had begun to fail. Examination revealed intense double papilloedema, and pupils which were rather large ($5\frac{1}{2}$ mm.) and immobile to light. There was no ocular paralysis at this stage, and though convergence was good the accompanying pupillary contraction was almost nil. A fortnight later associated upward movement of the eyes became very imperfect, and the pupils had come down to a diameter of $3\frac{1}{2}$ mm., the light reflex still being absent. Six weeks later upward movement was completely paralysed, and while the light reflex was lost both on direct and consensual stimulation the reaction on convergence was brisk, i.e., a double characteristic A.R. sign was obtained. Operation was offered, but, the patient's friends refusing consent, he was removed from hospital.

The importance of this case, apart from its identity with the preceding two as far as the combination of the A.R. pupil with tumours of the superior colliculi is concerned, resides in the proof it furnishes of variability under observation of the phenomenon itself and of the accompanying pupil changes. Thus the original stage of dilated and almost fixed pupils was followed by one of reduction in size coupled with the appearance of a typical A.R. sign, the former being associated with normal eye movements and the latter with paralysis of conjugate upward deviation. Since there is a common idea that in neurosyphilis the A.R. phenomenon, once developed, is a fixture, special attention must be drawn to the fluctuating nature of the dissociation in certain cases of mesencephalic lesion, a variability which is only to be expected, but which, at the same time, is of considerable pathogenic significance.

With one of my assistants (Dr. Rudolf) I¹³ have reported a highly characteristic case of mesencephalic tumour with double A.R. pupil and paralysis of vertical movement of the eyes. At autopsy a tumour apparently growing from the choroid plexus was found to have destroyed precisely the anterior corpora quadrigemina, whereas the posterior bodies and posterior part of the iter were intact.

Of interest in this connexion is a paper by Schuster¹⁴ on paralysis of vertical eye-movement. His second case is that of an old man of 62, with double A.R. pupil and paralysis of upward movement of the eyes, and with no other neurological sign. Post-mortem a

small softening was found in the mesencephalon, skirting the aqueduct on the right side, in the deeper layers of the tectum. The combination of A.R. pupil and paralysis of vertical movement, on which stress is laid above in tumour cases, is here exemplified by a vascular lesion in approximately the same area.

The occurrence, then, of the A.R. pupil in connexion with non-syphilitic lesions in the vicinity of aqueduct and anterior corpora quadrigemina is amply proved, and renders pointless the *ex cathedra* criticisms of Dunn,¹⁵ who declares that "it is impossible for me to understand how a tumour in the region of the third ventricle can produce a typical Argyll Robertson pupil. Every fact of the history of the growth of brain tumours and every fact of the history of the Argyll Robertson pupil pleads against such a possibility" (sic). His dogmatic assertions are discounted by his admission that he has never seen a case of mesencephalic tumour and by his uncompromising adherence to the view which assigns myosis an integral share in the syndrome.

Analogous instances from the literature are rather few and far between, but an excellent case is the old one of Moeli's¹⁶ (1887); in it a typical bilateral A.R. sign (pupils 5 mm. in diameter) resulted from a tumour of the third ventricle in a man of 57, vision being unaffected. From Weisenburg's¹⁷ useful paper on tumours of the third ventricle it would appear that "impaired pupil reactions" have been often observed, though the A.R. sign is not specifically mentioned. In this respect many of the cases quoted have been imperfectly examined. The reader may also be referred to Case 2 in a paper¹⁸ on *ectopia pupillæ* in mesencephalic lesions, where inequality of the pupils with sluggish reaction to light and brisk response on accommodation was caused by a colloid tumour of the third ventricle. Jelliffe and White¹⁹ state that the A.R. pupil has been found in cases both of third-ventricle and of pineal tumour, but do not give references. Farquhar Buzzard²⁰ has seen the phenomenon in non-syphilitic cases of mesencephalic tumour, those he briefly describes being for all practical purposes identical with my own in the combination of A.R. sign with paralysis of vertical movement of the eyes. To the objection of Cestan and Dupuy-Dutemps²¹ that sluggishness or fixity of the pupillary reflex to light in tumour cases is due to concomitant amaurosis the result of papilloedema, that such pupils are always dilated, and therefore that the condition is a "pseudo-Argyll Robertson sign," there is a ready answer; many cases of papilloedema are not

accompanied by any defect of vision. Vision was normal in Case 4 above, and but little diminished in Case 5, while in Case 6 the pupils were large and immobile to light at first and actually became smaller as the disease progressed and as vision became impaired. Moeli's case, too, disproves the contention.

5. Reflex iridoplegia, again, has been described in syringomyelia or syringobulbia (Lévi and Sauvinau,²² Dejerine and Mirallié,²³ Sicard and Galezowski,²⁴ Rose and Lemaitre,²⁵ and others). In Dejerine and Mirallié's case the A.R. phenomenon was unilateral, as in the case of Sicard and Galezowski, and in one of two published by Rose and Lemaitre. Interesting though these records are, their value would be increased were information as to the pathological lesions forthcoming.

Its occurrence in chronic alcoholism has been reported, among others, by Nonne,²⁶ whose case appears entirely free from criticism; in it Wassermann tests were repeatedly negative. One such case has come under personal observation.

CASE 7.—J. S., male, age 58, was under my care at the National Hospital for nervous symptoms associated with alcoholism. He had long been known to be a chronic tippler. I was called to see him at his own home one day, the history being that he had fallen in the street and had remained unconscious for almost twenty-four hours. On regaining consciousness he was confused and disoriented, and complained of his vision being "funny." When I examined him I found the optic discs clear. The pupils were rather small and immobile to bright light—on previous occasions their reactions had always been normal—both direct and consensual, whereas they reacted definitely on convergence. This movement of convergence, it should be said, was poor, and ocular movements generally were poor in range and not particularly well sustained in any direction. A diagnosis of polioencephalitis hæmorrhagica superior was made. After about six weeks, when the patient visited hospital again, both the light and the convergence movements of the pupils had much improved.

It is true that in this case a Wassermann test was not made, but apart from this regrettable omission I submit that the known facts in connexion with the case substantiate the clinical diagnosis.

Biermann²⁷ has put on record a good case of the A.R. pupil in diabetes mellitus, Wassermann tests being repeatedly negative. Mention may also be made of its development in the chronic hypertrophic interstitial neuritis of Dejerine-Sottas, and in some reported cases²⁸ of progressive muscular atrophy or amyotrophic lateral sclerosis. But no such cases can now be accepted as genuinely

non-syphilitic unless they have run the gauntlet of serological and spinal-fluid tests.

6. Of considerable interest and importance are the examples of the phenomenon of traumatic origin ; in fact, a surprising number have now been put on record, many of which are beyond cavil genuine A.R. cases.

Two groups may be distinguished : (a) those in which the lesion is in or behind the eye itself ; and (b) those in which the lesion is in the central nervous system. To the first of these belong the cases of Axenfeld,²⁹ in one of which the eye itself was injured by a splinter of wood ; with vision $\frac{6}{25}$, and a large pupil immobile to light, a prompt reaction on convergence was nevertheless obtained. Analogous examples from direct unilateral lesion of globe or orbit have been published by Cosmettatos,³⁰ Ohm,³¹ Velter,³² Abelsdorf,³³ and others. In Ohm's case a splinter of iron entering the eye was followed by mydriasis traumatica and fixity both for light and convergence ; several months later reaction on convergence returned, thus leaving a pure A.R. pupil, with good sight. Velter's case was that of an attempted suicide, with an orbital smash from a revolver bullet ; a unilateral A.R. pupil resulted. Other examples have followed fractures of the base (Axenfeld). In the second group are bilateral cases resulting from central traumatic lesions. Finkelnburg³⁴ has reported a case in an old man on whose head fell an iron instrument weighing some 150 lb., unconsciousness immediately resulting. In addition to general intracranial symptoms of concussion or contusion, the patient developed a bilateral A.R. phenomenon within a week or two of the accident, and the suggestion is it was due to minute hæmorrhages in peri-aqueductal grey matter, and consequent degeneration. A particularly interesting case is that recorded by Bergl.³⁵ The patient was a soldier concussed by the explosion of a shell twenty feet away ; in addition to familiar symptoms of intracranial commotio, within a few days a typical double A.R. pupil was found, the pupils being rather large, and yet after some three weeks of continuous observation the phenomenon disappeared and pupillary reactions became normal. Of other traumatic central cases that of Guillain, Rochon-Duvigneaud, and Troisier⁶ may be briefly mentioned.

The patient was a young man of 26 who had attempted suicide with a revolver, the bullet entering the right side of the neck just at the level of the hyoid, and being found by X-ray examination to have lodged in the

position of the right cerebral peduncle. A left hemiplegia ensued, with left hemianopia, some ptosis, weakness of some ocular muscles on the right, and a classical A.R. sign on the right and incomplete on the left (this pupil reacting sluggishly to light). Repeated examination of the spinal fluid for evidence of syphilis proved negative. It should be stated that the A.R. pupil developed under observation, for it was not present in typical form when the patient was admitted to hospital shortly after the injury.

I may finally cite the interesting case reported by Nattrass,³⁶ in which bilateral A.R. pupil and defective upward movement resulted from injury produced by a fragment of shrapnel which was shown to have traversed the upper part of the mesencephalon.

Further reference to these traumatic cases is made in a later paragraph.

From a consideration of the clinical material, personal and otherwise, at our disposal, pathological multiplicity for the A.R. phenomenon must be accepted, in which case questions as to its diagnostic and prognostic import assume a subsidiary position. Recourse to serological and spinal-fluid tests has taken the place of unsatisfactory speculations on the A.R. pupil as a criterion of active syphilis. It may undoubtedly remain as a neurosyphilitic "scar" long after active mischief has ceased, and cannot be held to be prognostic of anything. Its localizing importance, on the other hand, is fundamental, and is intimately bound up with the difficult question of its pathological physiology.

The Anatomo-physiological Arc for the Light Reflex

When light falls on the eye the physical stimulus sets in action two physiological mechanisms, one concerned with vision (the "sight" mechanism) and the other with reflex contraction of the pupil (the "light" mechanism). That these are physiologically distinct can admit of no doubt, though whether they are also anatomically separable is possibly not so certain, Magitot's³⁷ contention being that reflex-activating fibres are no more than collaterals of visual fibres, leaving the tract at a point near the external geniculate body. The following considerations, however, must be borne in mind.

1. The presence of both thick and thin fibres in optic nerve and tract, the former of which were traced by Monakow to the superior colliculi, but none of the latter, suggests a difference of function (substantiated also by Reichardt's³⁸ case), to which, further, the existence (according to Cajal³⁹) of single and multiple combinations

in the retina of ganglion cells with bipolar cells and rods or cones lends support; his plausible hypothesis is that the single combinations subserve vision and the multiple the light reflex.

2. In spite of the general truth that vision and reflex activity to light diminish *pari passu*, so that in complete optic atrophies the pupils are often immobile, exceptions are so numerous as to suggest an anatomical as well as the accepted physiological distinction between the two systems. In optic neuritis, optic atrophy, detachment of the retina, quinine amaurosis, etc., vision may be lost yet the pupillomotor reflex may persist; on the other hand, Axenfeld's and other peripheral traumatic cases show that with loss of the light reflex vision may persist. To assume an anatomical difference, represented also by difference in susceptibility or vulnerability, does not appear unjustifiable, although the subject could do well with less speculation and more patient anatomical research. In this connexion the case published by Reichardt³⁸ may be quoted, one of optic atrophy and amaurosis with conservation of the light reflex. Histological examination revealed persistence of a large number of undegenerated optic nerve fibres, whence it might be deduced that these were capable, during life, of mediating the light reflex but incapable of provoking a quantitative sensation of light (Magitot³⁷). But this interpretation (which is not Reichardt's) is at least doubtful, since the excellent researches of Léri⁴⁰ on tabetic optic atrophy have shown how few need be the persisting fibres in the optic nerve to allow of appreciation of light stimuli.

However this may be, a physiological differentiation for reflex impulses as opposed to visual impulses must be fully admitted; the physiological results both of stimulation and of destruction of the external geniculate body are different from those relating to the superior colliculi and prove the pupillary and visual paths are not identical.

THE REFLEX PUPILLOMOTOR PATH

Omitting in this place the question of the exact retinal origin of the light-reflex fibres, and leaving the visual route aside, we may proceed along the reflex pupillomotor path. It undergoes a partial crossing in the chiasma (analogous to the visual crossing), as is shown by the researches of Cajal and van Gehuchten. Parsons⁴¹ states that this partial decussation is also "proved by the hemiopic pupil reaction of Wernicke"; but less reliance can be placed on that reaction since the studies of Walker⁴² and others have demon-

strated its occurrence in some cases of posterior hemianopia, i.e., due to lesions behind the level of the external geniculate body, and an equally convincing case published by Dejerine and Jumentié,⁴³ the lesion being a hæmorrhagic softening which had destroyed inter alia the optic radiations posterior to the basal nuclei.

In the optic tract the pupillomotor reflex fibres can be followed to a point just before the external geniculate body is reached. The best method of physiological demonstration in this respect, that of constriction of the pupil by electrical stimulation of the optic nerve, has been utilized by Karplus and Kreidl,⁴⁴ who have found that all along the optic tract, except where it enters the lateral geniculate, electrical excitation contracts the pupil, and these pupil-controlling fibres can be followed, millimetre by millimetre, along the superior brachium (or "arm" of the corpus quadrigeminum anterius) and the anterolateral edge of the superior colliculus. At this point the electrical effect is again lost. From these important experimental results it may be taken that the pupillomotor reflex path avoids the external geniculate body and passes to the grey matter of the superior colliculus by the brachium. After bilateral section of the superior brachium the reaction of the pupils to light is absent; in this fashion Karplus and Kreidl have, in the ape, produced a bilateral A.R. sign which remained unchanged for eight months after the operation. They do not assert that in the A.R. phenomenon in man the lesion is of necessity at the same spot where they have been able to produce it experimentally, but their results are obviously of the first importance.

At this stage, however, it is desirable to deal with arguments denying to the superior colliculus and its brachium a place in the linked chain from retina to iris. Omitting the early experiments of Knoll (1869) and Bechterew (1884) we may consider the researches of Ferrier and Turner⁴⁵ (1901). These investigators destroyed with a cautery the region of the superior and inferior colliculi in seven monkeys; in all the experiments, with one exception, the pupils after the operation were contracted to the size of pin-points, but only temporarily, for after two days they returned to their natural size. In the authors' own words: "As regards the pupillary light reaction, *in the cases in which it was tested it appeared to be present (so far as reliance may be placed on this reaction when tested in animals, where it is so difficult to eliminate the complication of convergence and accommodation)*" (italics mine). To the evident hesitation of the authors themselves in the matter, further doubt

is added when it is remarked that a perusal of their protocols shows apparently that only *one* animal is recorded as having had its pupil reactions tested, and in it (see their Fig. 1) "the anterior border of the nates on the left side, and the posterior margin of the testes" remained. Thus their work can *not* be taken to prove that the whole anterior colliculus may be destroyed without preventing the light reflex, both because of the limited number of animals in which the pupillary reflexes were investigated and the admitted difficulties in the way of testing, and because in view of the uncertain path of the light reflex fibres from brachium to oculomotor nucleus no adequate proof is forthcoming from the experiments that these fibres could not have escaped. In fact, the complexity of cells and fibres in and beneath the superior colliculus is such that cases of incomplete lesion of that structure without apparent pupillary disorder cannot be taken to invalidate the views here advanced.

Special stress is laid on this matter, because Ferrier and Turner's experiments have been repeatedly taken to prove that the superior colliculi have nothing to do with the light reflex (Lhermitte,⁴⁶ for example, states that these observers "n'ont pu obtenir chez le chat ou le singe aucune modification de la réflectivité pupillaire par l'ablation de la paire antérieure des tubercules quadrijumeaux").

The experimental work of Levinsohn⁴⁷ must also be noticed. This observer destroyed *one* superior colliculus only in each of three apes, and after a few days found normal light reflexes in both eyes in them all. From photographs accompanying the paper it is not very clear precisely how much of the colliculus was destroyed; in one, at least, much of the peri-aqueductal grey matter seems to have been left intact. Nor is there mention of microscopical investigation of a sufficiently detailed character. Levinsohn does not state whether direct or consensual testing was made, and since only one colliculus was injured the experiments are open to some indefiniteness of interpretation.

Undoubtedly the crux of the whole question lies in the fact that the exact anatomical pathway from anterior colliculus to sympathetic nucleus in the oculomotor nuclear system has not yet been definitely traced, yet the difficulty resides not in poverty but in multiplicity of neuronal connexions. As Edinger⁴⁸ says, "Fasernetze und Züge, durch welche die Verbindung stattfinden könnte, sind in dieser Gegend mehrfach vorhanden. Das beweisende Experiment oder die beweisende klinische Beobachtung mit nachträglich erhobenem Befunde steht noch aus." It may be assumed

with a considerable degree of certainty that light-reflex neurones of retinal origin end at the level of brachium or superior colliculus—for after enucleation of the eyeball Probst ⁴⁹ has not been able to trace degeneration beyond the latter structure—and that there they enter into connexion with a new set. Of the various more or less alternating layers of grey and white matter in the superior colliculi, certain cells and arciform fibres therefrom derived, belonging to the deepest layer, engage our attention. They constitute the colliculonuclear tract, which takes origin especially in the large cells of the fourth layer and has both a direct and a crossed connexion with the mid-brain; the latter is made via the ‘fountain-like’ decussation of Meynert, whereby collaterals and terminal branches reach the oculomotor nucleus (and also the other ocular nuclei), while the uncrossed fibres descend to terminate in the homolateral third-nerve nucleus and in the others also (Tilney and Riley ⁵⁰). According to this description the crossed fibres skirt the aqueduct below to reach the opposite oculomotor cell-groups, but of no less importance is the dorsal crossing above the aqueduct, with subsequent connexion to the opposite nuclei, as already mentioned. This lamina commissuralis mesencephali (Edinger) is to be distinguished from the posterior commissure which lies in front of it, and with which it has been erroneously confused.

Paton and Mann,⁵¹ relying on the anatomical descriptions of Winkler,⁵² have recently suggested that the intercalated link between colliculus and oculomotor nucleus is constituted rather by the “fibræ radiales,” fine in calibre, stated by that authority to pass (from deep collicular cell-layers) more definitely to the neighbourhood of the Edinger-Westphal nucleus than the colliculonuclear tract. On this point further research is requisite.

Above, below, and laterally these fibres of the colliculonuclear system skirt the central grey matter of the aqueduct and would be the first to suffer from any peri-aqueductal toxic invasion. The significance of this statement is discussed more fully in a subsequent section.

In this fashion both the direct and the consensual reflex response of the pupil to light can be readily explained. If there is, as has been already stated, a partial decussation of pupillomotor reflex fibres in the chiasma, such a further semidecussation on the afferent side of the oculomotor nuclei must be conceded; otherwise the consensual reaction from the temporal side of the retina of the affected eye in a case of unilateral A.R. sign could not be explained,

as has been well pointed out by Harris.⁵³ The same author's study of the mechanism of the pupil reflex to light in animals and birds "proves the necessity for a posterior decussation of the pupil-reflex fibres between the corpus quadrigeminum or optic lobes and the third nuclei, total for those animals with total decussation of the optic nerves at the chiasma, and partial for those animals with semi-decussation of the optic nerves, in proportion to the size of the uncrossed bundle."

Though the experimental and clinical evidence now adduced argues strongly in favour of the passage of the light reflex by the superior colliculi, not a few writers have suggested a route which avoids these structures, viz., by the tractus peduncularis transversus, a fibre-system of uncertain origin and ending, which apparently leaves the optic tract at the outer side of the crus and adjoins the latter mesialwards to enter the mid-brain at the side of the emerging fibres of the third nerve. Here it appears to end in, or reach, a small nucleus first described by Bechterew and Marburg, which nucleus, according to Edinger, represents a part of the ciliary ganglion that has not wandered out to the orbit. While further research may enlighten us as to the anatomy and physiology of this tract, our present knowledge is insufficient to justify any speculation as to its possible connexion with the pupillomotor reflex.

Of other definite internuncial paths in the vicinity, reference may be made to the fasciculus longitudinalis dorsalis of Schütz, also known as the peri-ependymal longitudinal tract, consisting of a set of fine myelinated fibres immediately under, and above, the ependymal lining of the aqueduct, and therefore actually in the periaqueductal grey matter. Arising in a mesencephalic nucleus, the nucleus dorsalis tegmenti of Gudden, the tract seems to be of much less importance in man than in lower animals, since it represents a primitive motor pathway between olfactory lobe and ocular and lower cranial musculature. For anatomical and comparative anatomical reasons, notwithstanding its ocular connexions, this tract cannot be supposed to mediate the nerve impulses from superior colliculi to oculomotor nuclei, disorder of which is responsible for the A.R. pupil. Similar statements may be made of the familiar posterior longitudinal bundle—another and more important (partially descending) mesencephalic pathway; there is no clear anatomical proof of its conveying impulses from intercalated collicular neurones to intrinsic oculomotor nuclei. Finally, there remain the tectobulbar and tectospinal tracts, which arise in the

grey matter of the superior colliculi and constitute the outer section of the arciform fibres alluded to above as originating in the deeper cell-layers of the tectum. They cross below the aqueduct in the dorsal tegmental decussation of Meynert, between the twin red nuclei. The evidence suggests that these pathways serve to activate reflex movements in ocular, facial, neck, trunk, and arm musculatures in response to light stimuli via the anterior corpora quadrigemina, and, while the mechanisms are doubtless analogous to that responsible for the light reflex in the pupil, the latter is not carried out by these fibres (see below, however).

A further word of explanation is probably necessary. From what has been said already the reader will gather that the arciform fibres of the tectum are divisible into colliculonuclear, tectobulbar, and tectospinal sets, distinguishable, to some extent at least, by the levels at which they effect a decussation. It is possibly somewhat impracticable thus to differentiate three groups; at any rate, what has been called the colliculonuclear tract is included by some authorities in the tectobulbar system. Van Gehuchten,⁵⁴ for instance, states that tectobulbar fibres, in descending and crossing below the aqueduct to apply themselves to, though remaining distinct from, the posterior longitudinal bundle, give off collaterals to the cells of the third-nerve nucleus of both sides. Cajal, on the other hand, believes that the connexion between tectum and oculomotor nucleus is effected by his nucleus interstitialis, the origin, or one of the origins, of the posterior longitudinal fasciculus, collaterals from which, as is known, reach the third-nerve centres. We are, in fact, thus brought back to the view of Edinger, that the *possible* paths by which the junction can be brought about are abundant enough. It matters little, as a fact, what name is given to the innermost set of arciform fibres as long as we recognize that directly, or by the fountain decussation of Meynert or the dorsal decussation above the aqueduct, those of the fibres which are nearest to the aqueductal grey matter reach the homolateral and heterolateral oculomotor nuclei. Winkler considers the innermost set to be the fibræ radiales already mentioned, and claims they run *through* the grey matter round the aqueduct.

Perhaps the point may be made clearer from another aspect. The tectum opticum (optic lobes, region of the anterior corpora quadrigemina, superior or anterior colliculi, nates) is a reflex station of much significance and of wide relationships in connexion with light impressions, as is the region of the posterior corpora quadri-

gemina in connexion with auditory impressions. Taking only the former, it stands in anatomo-physiological affinity with homo- and heterolateral mesencephalo-ponto-bulbo-spinal centres for eye, face, head, neck, trunk, and limb movements in response to the stimulus of light. These are effected via descending crossed and uncrossed connexions represented by various tectofugal fibre-systems. Of these, the suggestion here advanced is that the most anterior is that concerned with reflex contraction of the pupil to light, the next anterior with movements of the eyeballs in response to light, and so on. For the moment, the physiological actuality of the most anterior connexion is of more importance for our purpose than the nomenclature we adopt for what appears to be the anatomical path for the reflex, viz., the fibres from the superior colliculi which border on the aqueductal grey matter above and below on their way to third-nerve centres.

THE IRIDOCONSTRICTOR CENTRE

Our next problem is to determine where among the latter is situated the iridoconstrictor centre, a problem, unfortunately, just as vexed as the other we have been discussing. To quote Parsons ⁴¹ again: "The number of reflex pupillary centres which have been described and localized by various writers, each with the utmost assurance, is bewildering in the extreme."

The iris-constricting centre is usually taken to be located in the nucleus of Edinger-Westphal, a small-celled paired nucleus well to the anterior end of the oculomotor group and close to the midline. Edinger ⁴⁸ himself believes from the cell-character of the nucleus (small, spindle-shaped or bipolar cells) that it is the visceromotor centre for the præganglionic fibres which, by a relay in the ciliary ganglion, are connected to postganglionic fibres running in the short ciliary nerves to the sphincter iridis; Westphal, ⁵⁵ finding the nucleus unchanged in a case of complete ophthalmoplegia externa, argued for its visceromotor nature by exclusion. Many objections, however, have been raised to this localization in the Edinger-Westphal nucleus of pupillomotor function: (1) According to Magitot ⁵⁶ the pupils react to light by the end of the fifth month of foetal development, but at that stage the cells of the Edinger-Westphal nuclei are completely undifferentiated, not being recognizable till the seventh month; only the ventral part of the principal lateral nucleus of the oculomotor constellation is then found to be recognizable. These statements, however, conflict with the recent studies of Paton and Mann ⁵¹; the latter conclude on developmental

grounds that the nucleus is concerned with pupil movements because its differentiation from the third-nerve nucleus "synchronizes with the development and functioning of the sphincter pupillæ musculature, both phylogenetically and ontogenetically."

(2) Magitot further remarks that the nucleus is not seen in any other animals than man and the ape; in fact, in the latter animal small-cell groups are scattered here and there in the oculomotor nucleus and central grey matter generally, "sicher sind sie aber nicht zu einem eigentlichen 'Kern' vereinigt" (Monakow⁵⁷).

(3) In several cases of fixity of the pupils to light Majano⁵⁸ found no pathological changes in the Edinger-Westphal nucleus, while in another case it showed outfall of cells though pupillary reactions were normal. Other observers have reported analogous cases. Frank⁵⁹ has adduced fresh pathological evidence to negative any connexion of the Edinger-Westphal nucleus with the iridoconstrictor mechanism; on the contrary, he considers his researches point to its being the mesencephalic centre for convergence, and with it he associates functionally the anterior median nucleus of the oculomotor group. As for the centre for iris-contraction to light, he thinks it may be found in the central grey matter dorsal to the nucleus of the fourth nerve, in the nucleus of Boettiger-Westphal, and with this he links the nucleus rapheos posterior, but the positive evidence brought forward in support of this localization is less impressive than the negative evidence in regard to the former. (4) Experiments by Bach⁶⁰ and Biervliet,⁶¹ and embryological researches by Tsuchida⁶² would appear to show that there is little specific localization either of individual external or internal muscles in definite parts of the oculomotor nucleus, but rather a diffuse or general localization. As far as the supply of non-striated muscles of the eye is concerned, Monakow concludes that their cells of origin are scattered mainly throughout the anterior (frontal) and mesial aspect of the principal lateral nuclei. Yet, if this were so, nuclear external ophthalmoplegias, the internal muscles unaffected, become more than ever difficult to explain.

Another by no means unattractive hypothesis may finally be mentioned. Is it possible that light-reflex fibres from the superior colliculi, in the colliculofugal pathway already described, do not actually enter the oculomotor nuclei, but join the third-nerve trunks immediately below the nuclei and run with them to the ciliary ganglia, where they end in relation to postganglionic fibres to the iris? This is the contention supported by Majano⁵⁸ with much carefully investigated clinicopathological and experimental

material. He has adduced evidence to show that the fasciculus longitudinalis prædorsalis, as it is sometimes termed—i.e., the tectobulbar tract, distinct from the posterior longitudinal bundle arising from the lateral nucleus of the superior colliculus and crossing in part in Meynert's dorsal decussation—contains fibres which run, homolaterally and heterolaterally, into third-nerve trunks directly and so to ciliary ganglia. In this fashion he simplifies the reflex arc considerably, reducing it to three components: (1) from retina to superior colliculus; (2) from colliculus to ciliary ganglion, a path which joins the oculomotor nerves but avoids their nuclei; (3) from ciliary ganglion to sphincter iridis. Uncertain though the matter is, evidence placing a pupilloconstrictor centre somewhere in the general oculomotor group is not to be lightly set aside.

It is known that visceromotor fibres of the third nerve are distinct from somatic-motor fibres, for in incomplete lesions of that nerve they may be intact when the latter are involved, and an old experiment of Schiff would appear to show that they are to be found to the inner side of the oculomotor trunk.

Of the last stage in the reflex arc, from ciliary ganglion to iris, little need be said. There can be no doubt that this ganglion is the actual peripheral sympathetic ganglion for the sphincter iridis, though the short ciliary nerves differ from other postganglionic fibres in that they are myelinated, i.e., they are white rami, and not grey. Excitation of the ganglion produces myosis; and destructive lesions, mydriasis to an all but maximum degree.

The Path for Convergence and Accommodation

The negative element in the A.R. sign being the failure of the pupil to respond to light, the positive element is its contraction with the effort of accommodation.

Ordinarily speaking, accommodation is a willed movement and therefore of cortical origin; in the act, three muscles take part—internal rectus, ciliary muscle, and sphincter iridis. Now the fundamental principle of cortical motor activity is that, in the cortex, movements and not muscles are represented. (But see also page 254.) The movement of closing the fist is cortical, and the cortex (it is thought) knows nothing of the muscles by which this movement is actually effected, viz., the three sets of flexors of fingers and thumb, extensors of carpus, and triceps, respectively. There is trustworthy evidence that the link between these groups takes place at the spinal level. Similarly, the movement of lateral

deviation of the eyes has its cortical centre at a known spot in front of the precentral gyrus, but the linking of homolateral internal rectus and heterolateral external rectus is effected at the pontine level by association between their respective nuclei. By analogy, we must hold that accommodation, as a movement, has a cortical centre, whereas the linking of its muscular components is peripheral, and is mediated by peripheral internuncial paths.

Of the actual site of this cortical centre we are ignorant, though, since the physiological centre for the oculorotary system (anatomically, the corticonuclear tract) is situated approximately at the junction of second frontal and precentral gyri, a twin accommodation centre, for convergence instead of lateral deviation, may conceivably lie in the same vicinity, corresponding areas in the two hemispheres being physiologically associated. Be this as it may, the corticonuclear tract proceeds by the internal capsule near the genu to the crus cerebri, whence the nuclear fibres pass dorsally by the pes lemnisci profundus, through the fillet, to the nuclei of the extrinsic ocular muscles; and it is no unwarranted speculation to suppose that by this tract also the impulse to effect accommodation in the triad of muscles already mentioned reaches their peripheral centre (perhaps the median nucleus of Perlia) in the oculomotor nucleus, from which, as far as the intrinsic muscles are concerned, the path must lie via the ciliary ganglion.

The exact relationship of the contraction of iris to ciliary-muscle and internal-rectus contractions respectively is immaterial for the purpose of this Chapter; suffice it to say that in the former we have an associated movement rather than a reflex in the strict sense, and that if this synkinesis is unusual in that it is a combination of somatic-motor and visceromotor elements, the fact that the sphincter iridis is phylogenetically a striate muscle should not be forgotten.

From this necessarily brief account of the anatomo-physiology of accommodation, the point of importance for the A.R. pupil that emerges is that distal to the oculomotor nucleus the pathways for iris-contraction in response to light and in association with convergence-accommodation are identical. Lesions of the oculomotor trunk never cause reflex iridoplegia; no claims for the existence of separate (sympathetic) fasciculi in the oculomotor trunk for the two visceromotor functions will stand investigation, though this is the contention made to explain some of the local traumatic cases (see below).

Site of the Lesion underlying the A.R. Sign

From the above descriptions it will be apparent that a unique or single localization for the A.R. sign is not to be expected, though clinical, experimental, and pathological evidence combines to place the lesion on the afferent side of the light-reflex arc, i.e., anywhere up to the synapse of pupillomotor reflex fibres with pupilloconstrictor centre in the third-nerve nucleus, or its vicinity.

1. In my view, by far the most common localization is in the neighbourhood of the aqueduct, where colliculonuclear fibres may be caught before they enter the oculomotor nucleus, and from which accommodation-fibres are sufficiently removed. Fresh evidence has been advanced in this Chapter of the association of the A.R. phenomenon with tumours of the superior colliculi and third ventricle, and on these cases I desire to lay the greatest stress. They prove beyond cavil the possibility of the development of the sign from non-syphilitic processes of mesencephalic origin on the afferent side of the third nuclear group. This view receives strong support from the researches of Karplus and Kreidl, which in reality provide that *experimentum crucis* hitherto awaiting.

No originality whatever is claimed for this view, since it has been advanced many times before, but the support it receives from the tumour cases given in detail above is sufficient to place it beyond the sphere of mere unverified hypothesis. Harris,⁵³ in a closely argued paper, stated that "although there is no positive evidence, I believe that the lesion of the Argyll Robertson pupil is a sclerosis of these fibres (fibres of Meynert's decussation), especially of their terminations in the neighbourhood of the third-nerve nuclei, a hypothesis which will account for all the phenomena of reflex iridoplegia."

There remains for some consideration the question of the frequency of the sign in neurosyphilis and its comparative rarity otherwise.

The neurologist is familiar with the peri-aqueductal degeneration often found in disseminated sclerosis; in fact, in that disease a subependymal sclerosis all round the ventricular system is common and one of its possible explanations is a toxic lymphogenous invasion from an infected cerebrospinal fluid (though some have argued for a special vascular arrangement). Now in syphilis and other infective states an ependymitis or subependymitis is no uncommon condition—e.g., the granular ependymitis of parenchymatous

neurosyphilis—and it appears to me a feasible speculation that there may be a special tendency for the syphilitic toxin to filter through to affect peri-aqueductal fibres, or terminal sensory arborizations, by lymphatic or possibly by vascular routes. A subependymal, peri-aqueductal, toxi-infective lymphatic spread will account for the frequency of the A.R. pupil in syphilis as readily as does a lymphogenous invasion via dorsal roots account for the absence of the knee-jerk. If it is asked why in such a postulated subependymal toxic spread the peri-aqueductal grey matter does not first suffer, one may legitimately point to the peculiar affinity of the syphilitic neurotoxin for afferent systems or for afferent terminal arborizations. In the search for a simple, uncomplicated, explanation of the *early* and *common* appearance of the A.R. pupil in neurosyphilis, at a time when the only other objective signs are likely to be toxic changes and evidence of meningeal reaction in the cerebrospinal fluid, I submit that the theory now advanced, of an irregular spread of the toxin through subependymal tissues surrounding the aqueduct to susceptible afferent fibres or terminal dendrites, will be found more feasible than any other.* The argument from subependymal changes in disseminated sclerosis (peri-aqueductal, periventricular, under fourth-ventricle floor, etc.) is topographically important though pathologically not to be stressed. By this theory the variations of the A.R. pupil in neurosyphilis—its uni- or bi-laterality, its absolute fixity or simple sluggishness, the presence or absence of consensual reflexes to light—can be readily understood. Jelliffe and White's¹⁹ view that "chronic meningeal exudates" in syphilis press upon the pupillomotor fibres in the brachium or anterolateral border of the superior colliculus is, I submit, incapable of explaining the early appearance of, and variations in, the A.R. phenomenon as satisfactorily as the one now offered. Moreover, early changes in the reflex arc being supposedly toxic, and not structural, one may in this way easily understand such observed facts as absolute A.R. pupils becoming relative, sluggish reactions becoming active, consensual reflexes returning, and so on.

2. In other and rarer cases, as we have seen, the site of the lesion is presumably nearer the back of the eye, in the course of the optic nerve or tract distal to the geniculate bodies. Axenfeld's unilateral traumatic cases can probably be elucidated on the supposition either

* Since this paper was first published (1921), in six cases of A.R. pupil definite thickening of the glia in the subependymal zone round the iter sylvii has been found by Warkany⁶³ (1924).

that sufficient fibres were left to allow visual, but not pupillomotor, impulses to pass, or that there is a difference of vulnerability between thick and thin fibres. The observers of some of the other local (orbital) traumatic cases do not, however, thus explain the phenomenon. Abelsdorf³³ thinks a partial lesion of the third-nerve trunk in his case will account for the reflex iridoplegia; Ohm,³¹ too, presupposes a double path in the nerve, one for the light-reflex and the other for the iris-reaction on convergence. These views are untenable, or at least unsupported by any pathological evidence. In cases of injury to the third-nerve trunk, Cushing⁶⁴ noted temporary paralysis of external muscles with intactness of internal muscles, but never a dissociated reflex in the latter.

3. Another theory assigns to lesions of the ciliary ganglion the phenomena of the A.R. pupil, a view urged notably by Marina⁶⁵ and Lafon,⁶⁶ among others. Marina found degeneration and chromatolysis of cells in the ciliary ganglia in neurosyphilitic cases, and supposed convergence-fibres run through the ganglion without interruption, only light-reflex fibres being relayed. But the manifold objections to this theory far outweigh its possible attractiveness. The results of experimental paralysis of the ganglion (dilatation and complete immobility of the pupil for all stimuli) are utterly different from the clinical features of the A.R. sign; ganglionic lesions will not explain the myosis of many A.R. pupils, or the conservation of consensual reflexes in many instances; and not readily the absence of the dilatation-reflex following painful excitation. Pathologically, Thomas⁶⁷ found no degeneration in short ciliary nerves, ciliary ganglionic cells, or in proximal roots of the ciliary ganglion, in three cases of the A.R. phenomenon in *tabes dorsalis*. Lafon's attempt to override the manifest difficulties by supposing that syphilitic invasion of the ganglia causes "perversion" and not "paralysis" of function, is, as it seems to me, almost a *petitio principii*.

In his Lumleian Lectures, Sir David Ferrier⁶⁸ has upheld the ciliary-ganglion theory nevertheless, although he apparently adopts it to explain, not all cases of A.R. pupil, but only those of neurosyphilis. The endeavour is made to account for the dissociated reflex, not by Marina's view that fibres for convergence-accommodation pass through the ganglion without interruption, but by supposing that "different nerve-fibres in the same trunk may be differently affected by destructive or toxic agencies"; the speculation is that syphilis may so affect the ciliary ganglion and

ciliary nerves that though these "cannot transmit the reflex impulse of light to the sphincter pupillæ, they can readily allow the more powerful stimulus associated with accommodation to pass through." Why the assumption of a more powerful stimulus for accommodation should be made is not very clear, nor is the grave objection met that the theory fails to explain the preservation of the consensual light reflex in numerous A.R. cases. Besides, as already noted, the ciliary ganglion and short ciliaries have been found to be normal precisely in some cases of tabes with A.R. pupil—a serious blow to the theory.

It ought, perhaps, to be stated that changes in the ciliary ganglion in neurosyphilis are in no way excluded as an occasional or even a frequent occurrence, and with them the occasional or frequent irregularity of the syphilitic pupil may possibly be associated. But it must at the same time be pointed out that the argument (for the ciliary origin of the A.R. sign) which depends on a presumed association between pupil irregularity and the subsequent development of the A.R. phenomenon loses force in view of the fact that a central (mesencephalic) origin for some pupil irregularities must be admitted ¹⁸ (cf. Case 4 above).

The Myosis of many A.R. Pupils

Though in numerous instances the A.R. phenomenon is observed in pupils of normal dimensions, the fact remains that in some 30 per cent., according to Uhthoff (quoted by Lutz ⁶⁹), myosis accompanies it. Some of the proposed explanations of this concomitant myosis are rather vague. Higier ⁷⁰ says it is due to "absence of sensory stimuli owing to disease of the posterior columns of the cord"; Argyll Robertson ¹ himself stated that "for contraction of the pupil under light it is necessary that the ciliospinal nerves remain intact, and as in these cases of myosis the ciliospinal nerves are paralysed, light does not influence the pupil." But the spinal explanation of myosis, possible enough though it be in some instances, will not serve for all by any means, least of all for myosis accompanying the A.R. sign.

The myosis of the A.R. pupil cannot be due to irritation or excitation, for it may continue for years, and is unaccompanied by any spasm of accommodation (not that the latter point always signifies). Ferrier's ⁶⁸ opinion that it is caused by "degenerative changes of an irritative character in the sphincter itself" is open to the same objection; how irritation can be caused by degeneration in a

minute muscle and continue indefinitely is difficult to conceive. The myosis must be the sequel, in large part at least, to paralysis of the pupillodilator mechanism, for which the descending sympathetic path to the so-called ciliospinal centre of Budge, thence via the superior cervical sympathetic ganglion to the carotid plexus, Gasserian ganglion, ophthalmic division, and long ciliary nerves to the iris, is well known. Some iridodilator fibres, however, probably pass from the carotid plexus to the sympathetic root of the ciliary ganglion and so possibly to the iris, for after gasserectomy the pupil never remains permanently contracted. A paralytic lesion of the descending pathway will occasion myosis without the A.R. phenomenon; for the combination, we must seek a solution at higher levels.

The descending iridodilator tract just described is known at the level of the medulla, where occluding lesions of the posterior inferior cerebellar artery cause softening of the lateral aspect of the medulla, one of the symptoms of which is homolateral myosis. Evidently, then, the pathway lies somewhere in the *formatio reticularis*—according to some, near the *fasciculus solitarius*. Higher up still, Spiller⁷¹ has in two cases of pontine lesion found evidence of involvement of the tract in the shape of homolateral myosis, deducing therefrom absence of decussation below that level. Here, clinico-pathological evidence comes to an end, but, fortunately, at the same point experimental work comes to our assistance. At a spot on the base, lateral to the infundibulum, near the exit of the third nerve, and just behind the optic tract, Karplus and Kreidl⁷² have in a series of twenty cats obtained with constancy, on electrical stimulation, a maximum dilatation of both pupils; and have shown by suitable procedures that this stimulus is definitely transmitted by the homolateral peduncle, and crosses in part at a lower level, to descend to the cervical cord and so back to the eye by the familiar route. Further research by the same observers points to the sympathetic centre for this iridodilator path being located in the *regio subthalamica*, dorsomesial to the *pes pedunculi*, in the frontal part of the *corpus Luysii*.

Though this experimental work requires verification we might be able to explain the frequent combination of A.R. pupil and myosis on the assumption that the iridodilating tract on its way through the mesencephalon passes near Meynert's dorsal tegmental decussation or near the aqueduct, but this is at present purely speculative. Lutz's⁶⁹ hypothesis is that the descending fibres run

in the tectospinal tract, and that lesions of the tectobulbar tract cause the light immobility, and of the tectospinal tract the myosis. The suggestion is interesting, but equally speculative, though by exclusion it is easy to demonstrate that the myosis of some A.R. pupils cannot result from involvement of the third-nerve sector and must be the outcome of a more centrally situated lesion. The problem, simply stated, is: Where do the light-reflex arc and the pupillodilator path come sufficiently close to each other to be simultaneously implicated in a common destructive lesion? In the present state of our knowledge the question is more easily posed than answered.

There is reason to believe that the effect of the pupillodilator mechanism is double; i.e., the iridodilator centre of the mesencephalon (or wherever exactly it be) exercises, in functional activity, both a stimulating action on the iris via the spinal route and an inhibiting action on the iridoconstrictor centre of the third-nerve nucleus by another—direct—route to the latter. Should, then, the dilating mechanism be at fault, or interfered with, the pupil contracts doubly, as it were, because of the loss of this inhibiting influence on the tonus of the constrictor centre.

Another interpretation of the myosis of the A.R. pupil is nevertheless possible, since the constrictor tonus of the centre in the third-nerve group is modifiable from another direction.

It is known, and has been experimentally demonstrated,⁴¹ that dilatation of the pupils can occur when the usual dilator tract is out of action; the explanation offered, also supported by experimental evidence, is that the tonus of the pupilloconstrictor centre in the oculomotor group can be directly inhibited from the cortex. Though the paths by which this inhibitory influence is exercised are unknown there is a very definite anatomical connexion between cortex and corpora quadrigemina in the form of the palliotectal system of fibres, which end in large measure in the superior colliculus. The assumption is warrantable, in fact probable, that the physiological influence passes thence by peri-aqueductal lines to the iris-constricting centre in the oculomotor group. Assuming that among these are the fibres stimulation of which inhibits the tonus of the latter centre, causing dilatation, then we must, conversely, consider that paralysis of these same fibres removes this cortical inhibitory action, hence overaction of the third-nerve constrictor effects, and consequent myosis.

Now it will be at once apparent that this hypothesis fits in with

the view which assigns to destructive lesions in the neighbourhood of the aqueduct the usual appearance of the A.R. phenomenon, so that in this fashion the frequent myosis may find a simple explanation. Not only so; by such a postulated mechanism the common observation that the A.R. pupil does not respond to painful or emotional stimuli by dilatation can be readily understood. Such suitable excitations as should lead, by this psychoreflex mechanism, to inhibition of the tonus of the third constrictor centre or centres, and consequent dilatation, fail to reach the latter owing to the assumed interruption of functional activity between the pallio-ectal terminations in the superior colliculi and the oculomotor group.

Few problems in organic neurology will so well repay further minute anatomical and physiological research as the A.R. phenomenon, which has been a *Haupttummelplatz* of controversy for the last fifty years, but is within reach of a final solution.

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