Observations on the physiology and pathology of hemi-chorea / by J. Hughlings-Jackson, M.D.

OBSERVATIONS

ON

THE PHYSIOLOGY AND PATHOLOGY

OF

HEMI-CHOREA.

BY

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PHYSIOLOGY AND PATHOLOGY OF HEMI-CHOREA.

I HERE bring together, and as far as possible add to, the arguments I have already in many places advanced bearing on the speculations I put forward several years ago, on the Physiology and Pathology of Chorea. This article necessarily therefore involves very considerable recapitulations.[1]

It has long seemed to me that embolism (see p. 8) of parts in the region of the corpus striatum gives a most satisfactory explanation of the physiology and pathology of cases of chorea. This view has found very few supporters. Dr Broadbent, however, arrived at similar conclusions quite independently and almost contemporaneously. In a paper read before the Medical Society of London, session 1865–6, he tried to show that chorea must be owing to an affection of the corpus striatum or thalamus opticus. Dr Broadbent referred to my views, and agreed with me generally as to the seat and nature of the changes in this disease. A year or two ago, Dr Barnes read, before the Hunterian Society, an able paper on "Chorea in Pregnancy." In the discussion which followed I advanced the views which I state in this paper. Only one speaker, and he in a very limited way, countenanced my notions. One physician, whose name would carry the greatest weight, said that he did not see the slightest reason in support of such a conclusion.

Very recently, June 1868, Dr Barnes has once more brought the question before the profession, in an important paper read before the Obstetrical Society of London. He objected very strongly to the views I hold. Again, only one of the speakers in the discussion agreed with me as to the nature of the changes in chorea, and he very generally; and no one agreed with me as to their position. The following extracts from the report given in the medical journals[2] can of course convey but an imperfect idea of Dr Barnes's admirable paper:—

"The author subjected to analysis the theory that the proximate cause of chorea was embolism in the capillaries of the brain, derived from vegetations on the cardiac valves, the consequence of rheumatism,—a theory maintained by Kirkes, Hughlings-Jackson, Tuck-

[1] See Lond. Hosp. Reports, vol. i. 1864, p. 459. Lancet, Nov. 26, 1864, p. 606. Med. Times and Gazette, Jan. 28, 1865; Dec. 14 and 21, 1867; and Aug. 1, 1868, p. 137. Roy. Lond. Ophth. Hosp. Rep., Dec. 1866, p. 287. St Andrew's Reports, vol. i.

[2] See Med. Times and Gazette, Aug. 1, 1868, p. 137.

well, and others. He urged clinical facts as inconsistent with this theory, at least in the exclusive and absolute form." . . .

"The author put forward the speculation that the chorea at first was due to functional irritation of the brain, and that the softening and decided lesions found in some fatal cases were the result of the chorea, the consequence of altered nutrition caused by the repeated shocks and waste of nerve-force attending the chorea."

My friend and colleague, Dr C. B. Radcliffe, in a footnote to his valuable article on Chorea (*Reynolds's System of Medicine*, vol. ii. p. 127), refers to the views I hold on the physiology and pathology of this disease in a manner at which I cannot be otherwise than highly gratified; but he adds, "As yet the clinical facts are wanting which are necessary to give the theory a firm foundation." I am the more sorry for this conclusion, as I think the clinical facts form the strongest defence of the position I hold.

Recently, Dr Tuckwell of Oxford has written an important paper on chorea (*Med.-Chir. Review*, Oct. 1867). Dr Tuckwell thinks that embolism is the usual cause of this disease. In the next number of the *Review* Dr John W. Ogle brought forward a large number of cases of chorea, but he did not endorse the theory of embolism. In the *Med. Times and Gazette*, May 30, 1868, will be found an article full of clinical facts, by Dr Russell of Birmingham, in which he upholds the main views stated in this paper.

There are three chief divisions of the hypothesis. I shall illustrate by cases of hemi-chorea, although of course what follows, *mutatis mutandis*, will apply to cases of bilateral chorea:—

1*st*, That, in chorea, nerve-tissue forming the convolutions near to the corpus striatum, is in parts diseased.[1]

2*d*, That the nerve-tissue there diseased is *not* destroyed, but is unstable.

3*d*, That this local instability is frequently brought about by under-nutrition consequent on diminished supply of blood—anæmia—caused by blocking of *small* branches of the arteries supplying the convolutions referred to. I will state what I have to say under the following heads:—

1. The muscular *Region* affected.
2. The *Nature* of the movements produced.

[1] The following is of much interest with regard to this point:—"An interesting investigation of the brain in a case of general chorea has been recorded by Dr Aitken in the Glasgow Medical Journal, No. 1, 1853. He noted the important fact that the specific gravity of the corpus striatum and optic thalamus on the right side was 1025, and that of the same parts on the left side was 1031. Further observations on this subject are greatly needed, and if made with the same minuteness and exactness as those of Dr Aitken, will no doubt throw great light on the pathology of chorea and other allied affections."—*Todd's Lectures on Nervous Disease.*

Although I think the affection is of the convolutions (see p. 6), I shall be satisfied with proving the first step—that the morbid changes in chorea are in the region of the higher motor tract. Indeed (see p. 11) I chiefly illustrate by the corpus striatum alone.

3. The local *Internal* change.
4. The *Time* of the movements.

1. The Muscular Region affected.

It is, I take it for granted, the external muscular region affected, which points to the internal nervous organ or part damaged. The following facts supplied by cases of hemi-chorea point, I submit, most strongly towards the locality of the corpus striatum, which is the part damaged in the common form of hemiplegia.

(*a.*) The movements are often one-sided. Dr Todd is very decided on this point. I have prepared a series of quotations from his works, but I do not print them, as I find that an authority of equal reputation, Trousseau, thinks that *hemi*-chorea is rare.

For my own part, I have no doubt at all that hemi-chorea is common. Moreover, when chorea is bilateral, one side is usually *more* affected than the other. The quasi-accidental process of embolism is the very one likely to produce sometimes unilateral, sometimes bilateral, sometimes symptoms unequal on the two sides. Russell says, "Now, I have to add that, so far as my observation has enabled me to judge, even although the chorea may be ultimately bilateral, it *commences unilaterally* in a large majority of instances, and remains unilateral for a distinct, and sometimes a considerable period of time.

(*b.*) The side affected is (as in hemiplegia from embolism) generally the right side. It is, however, proper to say that Austin Flint and Trousseau hold that it is usually the *left*. In Russell's cases, right 18, left 11.

(*c.*) The muscles most moved in hemi-chorea are those most palsied in hemiplegia from destruction of part of the corpus striatum. In these so-called one-sided cases, however, the trunk-muscles—or rather the muscles acting bilaterally—are involved, and they are *not* involved in hemiplegia. It is for this reason only that I add the word *most*. This is now, thanks to Dr Broadbent's highly important hypothesis on unilateral and bilateral movements, not the difficulty that appears.[1] Broadbent's hypothesis accounts most satisfactorily for the seeming discrepancy that the bilateral muscles are involved in hemi-chorea and hemi-spasm, whilst they escape in hemiplegia. Indeed, I think I have, from observation of cases of hemi-spasm, verified Dr Broadbent's hypothesis.[2]

It is plain that the disease *must* be above the spinal cord when the face is involved. Besides, hemi-chorea has no counterpart in the permanent effects of lesion of one lateral half of the spinal cord. The disease is scarcely likely to be in the pons Varolii, as the face is always involved on the *same* side as the limbs. There are, however, seeming discrepancies here, but these also Broadbent's hypothesis will resolve. If the lesion were in the pons, the face

[1] See Med.-Chir. Review, April 1866; Ranking's Abstract, 1866.
[2] See Lancet, vol. i., 1867, and Med. Times and Gazette, Aug. 15, 1868.

ought *sometimes* to be affected on the side opposite to the irregular moving arm and leg.

(*d.*) As in most cases of hemiplegia, the leg suffers less than the arm.[1] This is the case, too, with hemi-spasm, although there are exceptional cases.[2]

(*e.*) There is usually greater defect of speech when the right side is affected.[3] However, when the left side is apparently solely affected, speech is sometimes disordered. The reason is obvious.

(*f.*) The movements sometimes die away into hemiplegia quite like that which destruction of nerve-tissue in the corpus striatum produces, and in almost all cases there is *some* paralysis. Occasionally, but yet rarely, we find the paralysis almost as absolute in degree as that found in hemiplegia from clot.

The foregoing evidence points, I submit, to the region of the corpus striatum. It is convenient, although seemingly out of order, to consider next the *condition* of the muscles in hemi-chorea.

2. The Nature of the Movements.

It is not denied that "disorderly movements" occur with disease in many parts, probably in most parts, of the nervous system. What I wish to show is, that *certain* irregular movements—often affecting the face, arm, and leg of one side only—occur from disease of the higher centres of movement, viz., of convolutions on the hemispherical side of the corpus striatum. It is especially to be observed that they differ from the jerky movements of the arm occasionally seen in severe cases of locomotor ataxy. They are not mere spasms and cramps, but an aimless profusion of movements of considerable complexity, much nearer the purposive movements of health. They are not so much incoherences of muscles (like the "fist" we see in a partial fit of those convulsions which begin unilaterally where all the muscles of the hand are in action at once) as incoherences of *movements of muscles.* There is some method in their madness. They are not analogous to playing at once many keys of a piano in mere order of continuity, but to a random playing of harmonious chords. Again, they are *successions* of movements; moreover, they are successions of *different* movements. Now, it is clear that close upon the corpus striatum lie the rudimentary arrangements of fibres and cells for the highly complex and widely associated movements of speech; and it is, I think, independently of other arguments, at least plausible that corresponding movements of the arm—which may be called, according to our stand-point of thought, either rudimentary psychical, or highly developed physical movements—should have their "centres" here too.

To guard against misunderstanding, I must say that I do not hold that functions have abrupt geographical localizations. I

[1] On this point see Russell, Med. Times and Gazette, May 30, 1868.
[2] See Med. Times and Gazette, Aug. 15, 1868, p. 178.
[3] See a case of mine reported, Lancet, Nov. 26, 1864.

suppose that the rudimentary psychical movements of speech, and the corresponding movements of the limbs, are represented in *each* part near the corpus striatum, although I think it likely that the former are specially represented towards the anterior and the latter towards the posterior lobe.

Although I have instanced series of comparatively simple movements of single parts, analogous to ataxy of articulation, there are, in some cases of chorea, movements of much wider range, implying, I imagine, changes deeper and wider in the hemisphere; just as there are movements (*i.e.*, misuse of words or incoherence) dependent, doubtless, on changes further spread in the (left) hemisphere than the parts close to the corpus striatum. Saying nothing of the acutest cases, the Mental, or, as it is usually called, the Emotional condition of most choreal patients countenances the same notion. There is less evidence on the point; but I think it likely that puerperal mania results from instability of larger and more distant parts of the hemisphere usually produced in a similar manner. But we must begin our studies of mind by a consideration of the more rudimentary phenomena, although we do not make arbitrary distinctions betwixt those which are grossly motor and those sensori-motor impulses which we speak of as being mental. I shall therefore continue to speak of hemi-chorea.

3. The Local Internal Change.

I shall consider its (*a.*) *General* nature; (*b.*) its *Particular* nature, as a tissue-change; and, under (4.), what I may call its (*c.*) *Abstract*[1] nature.

(*a.*) *The General Nature of the Local Change.*—The fact that we have never agreed as to its nature, is some evidence that the change in nervous tissue in chorea is a *minute* one. Austin Flint says, "As one of the neuroses, this affection, of course, has no anatomical characters." A valuable negative argument is that, although patients who are the subjects of chorea may complain of headache, they never, or certainly very rarely, suffer the intense pain in the head or urgent vomiting which occurs with *coarse* disease of the brain, and, to the best of my knowledge, there are never any very great changes—usually none—in the optic discs. In this I am glad to be able to say that Dr Clifford Allbutt agrees with me.

From a superficial point of view, it is, I think, most striking, that great defect of sight (from optic neuritis) occurs not unfrequently with unilateral palsy and with unilateral spasm, but *never* with unilateral chorea. The real fact is, that optic neuritis occurs with *coarse disease* of the brain.[2] It is true, and indeed very significant, that from coarse disease of one cerebral hemisphere, we may have almost all sorts of conditions of muscles of the arm. But

[1] "Abstractness means *detachment from* the incidents of particular cases. Generality means *manifestation in* numerous cases."—*Herbert Spencer.*

[2] See Med. Times and Gazette, Aug. 15, 1868, p. 178.

choreiform movements are rare, and they are not quite like those seen in choreaic children, and in most cases they are only developed when the patient voluntarily uses the affected limbs.

(*b.*) *The Particular Nature of the Local Change.*—The speculation advanced, as to the supposed particular nature of the local change, is in harmony with (*a.*).

It is admitted by all that we find in chorea, at least frequently, a *condition for* embolism. In some cases there is valvular murmur. In others there is a history of rheumatic or of scarlet fever. The movements will sometimes begin with joint affection; this being either what would be commonly called acute rheumatism, or slight pain and swelling in a few joints. Add to this that, according to Kirkes, Wilks, and Andrew, vegetations are *invariably* found on the cardiac valves at post-mortem examinations on those who have died of chorea. I am aware, however, that there have been exceptions. Dr Kirkes long ago put forward the view that chorea is associated not so much with rheumatism as with valvular disease of the heart, a thing which rheumatism often induces. Then chorea occurs with pregnancy and parturition, states under which it is admitted that *paralysis* from embolism results.

To complete what I have to say on this part of the question, I will quote one of our best clinical observers.

Dr Hillier, in his recently published work on "Diseases of Children" (1868), writes:—"The late Dr Kirkes, in one of his last papers (*Medical Times and Gazette*, June 1863), proposes a very ingenious and plausible theory to explain this connexion. He dwells upon the frequent presence of a systolic murmur in chorea, and states that valvular disease often exists without the presence of a murmur to indicate its existence, especially when granulations occur on the auricular surface of the flaps of the mitral or tricuspid valve. He also calls attention to the fact that a condition, which has not been uncommonly found post-mortem in fatal cases of chorea, is softening of parts of the cord or of the cerebrum, and occasionally intense capillary congestion of other parts, more or less simulating hæmorrhage. The patients who suffer from chorea are very impressible and emotional, and very liable to derangement of the nervous system. Putting these facts together, he asks, may not endocarditis be the first link in the chain, causing fibrinous exudation on the cardiac valves? Particles of this exudation are carried forward into the smaller vessels of the cord and brain, giving rise to deranged function in these parts, and sometimes leading to local softening of nerve-tissue." However, Dr Hillier adds—the italics are mine:—"*A forcible objection to this theory is, that we do not find symptoms like those of chorea produced by embolism, when it is proved to exist, or when experimentally induced in animals.*"

I do not consider this argument to be so forcible as it appears at first glance.

4. The Time of the Movements.

The symptoms which *Destruction* of nerve-tissue in the corpus striatum produces are well known, as this part of the nervous system is frequently much damaged, and the symptoms resulting are obtrusive and often permanent. I fear, however, that we do not seek with sufficient intentness the disorders of function which correspond to such losses of function.[1] In other words, we do not study muscular disorders as defects of co-ordination in Time.

Just as *loss* of function, for instance palsy, follows *destruction* of nerve-tissue, *however produced*—by clot, by tumour, by injury, etc. —so *disorder* of function, for instance chorea or spasm, results from *instability* of nerve-tissue, *however produced*—by mechanically-produced anæmia, or by inflammatory changes in the imbedding tissues, and, as I think, by embolism.

We have to think not only of changes in nerve-tissue of various *quantities* decided enough to produce different *quantities* of *loss* of function—such as degrees of impaired power of movement from weakness to absolute palsy—but also of degrees of *quality* of change allowing degrees of disorderly function, such as movements in incoherent succession. This distinction is, I think, neglected in our study of cases of so-called Aphasia.[2]

After this necessary digression, I return to consider a special case. Still keeping to the simplest cases, those of *unilateral* chorea, such as we frequently see in the out-patient room, when in a boy or girl, who has valvular disease of the heart, a certain muscular region, viz., that governed by the corpus striatum, becomes choreaic – we may ask, why *this disorder* of movement is not ascribed to embolism, as the *corresponding palsy* of movement would certainly be?

It has been urged that we can well understand how, from blocking of bloodvessels supplying a motor centre, there might follow *palsy*, but not "activity" of the muscles it governs. To this I would say, in the first place, that even if a *small part of the corpus striatum were actually destroyed* as by a clot, or if a small part of the convolutions near it—near the left one at least—were practically wanting, there *need* be no permanent motor symptoms of any sort. Of course, too, if an equivalent of either were partly destroyed from anæmia, the result of a mechanical block, the result would be the same.

If a *large* quantity were *destroyed* there would then be motor

[1] The word function has a threefold meaning when used in speaking of the healthy nervous system. (1.) It is the function of *nervous Matter* to store up force for *future* expenditure. (2.) It is the function of nerve Units, in expending their stored-up force, to develop *certain* orderly, and more or less complex, movements. (3.) It is the function of nervous matter, forming the nerve-units of particular Organs, to expend their stored-up force in developing certain more or less complex movements in *correspondence with* special—not always constant —excitations, which bring the local movements into harmony with the whole organism.

[2] See Med. Times and Gazette, June 23, 1866; Aug. 15, 1868.

symptoms; but *these would be those of loss of function—palsy.* There could not be *disorder* of function—such as irregular movements—from *destruction* of nerve-tissue; to assert that there could be, is equivalent to saying that there could be disorderly function of parts non-existent. But I do not suppose either that a large quantity of the corpus striatum region is involved in the embolic anæmia, or that the small part which is involved, and on which the irregular movements depend, is sufficiently ill-nourished to lead to *loss* of its function. In other words, in the giving rise to choreal movements, as we usually see them, but little nerve-tissue is affected, and that little is not destroyed.

For it must be admitted, that when a group of muscles is immovable from a diseased change in the nerve-cells and fibres which govern that group, those cells and fibres are practically destroyed, so long as the muscles remain utterly incapable. But if the same muscles are copiously moving, as in chorea, it is plain that their governing cells and fibres cannot be destroyed. Again, since the movements are disorderly in Association and in Succession, the nerve-cells and fibres cannot be healthy. To repeat, wherever in cases of chorea the change may be, and whatever may be its *particular* nature, it is clear that its *abstract* nature must be one betwixt loss of function, leading to paralysis of motion, and healthy function allowing the orderly movements of health. In short, nerve-tissue in chorea is neither destroyed nor is it healthy—it is unstable. This is again in accordance with the fact that the change is a minute one. We can scarcely expect to discover[1] in nerve-cells and fibres the very changes on which a disorderly exaggeration of the normal function of a nervous organ depends.

It may, however, be urged that there should be *some* paralysis, if there be a *local* change of any sort. There usually is; but this I presume depends on more decided changes, such as will be equivalent to destruction of nerve-tissue. Still, there are cases of which it might plausibly be said that there is at least no obvious palsy.

When a sufficient quantity of *any* part of the corpus striatum is destroyed, the arm is permanently weakened or completely paralyzed. But it must not be overlooked that the limb is "partially paralyzed;" not paralyzed in some one or more parts only, although one part of the limb may suffer more than the rest. From this we may infer that the muscular system of the arm—the *whole* of the arm movements—is represented in *each* part of the corpus striatum; and thus I account for the fact before stated (p. 9), that permanent loss of a small part of this motor centre may be borne without palsy, or that the palsy may be recovered from.

[1] Although we may not discover the change in *nerve-tissue* which allows disorderly movements of muscles, which in health that nerve-tissue governs in an orderly manner, we may find changes in the other ingredients of the nervous *organ*. Besides, there is in chorea usually some palsy of movement, and thus the parts corresponding may be obviously changed.

In other words, using a term borrowed from Spencer, the corpus striatum is a series of Physiological Units—each unit representing not some one *part of the limb*, but potentially the *whole* of the limb. (I have entered further into this point as regards Localization of "Faculties,"—*Med. Times and Gazette*, Dec. 14 and 21, 1867, Aug. 15 and 22, 1868; *London Hosp. Reports*, vol. ii. p. 238; and *Roy. Lond. Ophth. Hosp. Rep.*, Dec. 1866.) Still these nerve-units are not supposed to be mere repetitions one of another, but each is supposed to represent some special *leading* movement of the *whole* limb.

But although *destruction* of a *small* part of the corpus striatum need not produce any symptoms, or at least any permanent symptoms, it is plain that *instability* of a small part *must* result in symptoms, and it is equally plain that the whole of the arm *must* suffer when *any* considerable part of the corpus striatum is unstable.

Thus becomes more simple the apparent paradox, that with extensive gross disease of the cerebrum or cerebellum (for instance an abscess) *destroying* a large tract of nerve-tissue, there are sometimes no obvious motor or mental symptoms; and that, on the other hand, when a patient dies insane, or of disease so dramatic as acute chorea, we cannot discover the morbid change which is to blame for the symptoms.

The word instability has been objected to. Its use is, of course, a mere verbal artifice. It names a condition of nerve-tissue of which none of us have a clear notion. It is pretty much like "excitability," a word freely used in a similar way. But "instability," as I use the word, differs from "excitability," as the latter is generally used. Instability is supposed to be caused by *defective* nutrition. The "exaggeration" of normal function depends, I suppose, on the changes being permanently spread over a wider area than the temporary changes which allow the usual orderly movements of health.

There is one thing which is not easily accounted for; namely, how it comes to pass that chorea rarely occurs in infants and in people of middle life. It may be said that in infants there is not so often a condition for embolism.

Since—at least so it seems to me—the disorder in chorea is of movements which are acquired, and which are probably only fully learned by a very long apprenticeship, I used to suppose that the nervous arrangements for these movements were but partially developed in children, and that the motor processes involved, and their arterial integration, would be, so to speak, caught in a stage of incomplete development—in short, that these centres were diseased when *half*-educated. However, movements very like—I think quite like—those of chorea in children, do occur, but yet most rarely, in infants and in old people.

I have used above the term *arterial integration*. My speculation is, that the cerebral arteries in health have to do with the *orderly development* of the functions of nervous centres, and that their con-

traction is one factor in the *disorderly development* of the phenomena of chorea, convulsion, etc.,[1] the quasi-permanent, internal local instability (see p. 10) being the other factor.

It is admitted that the irregular movements often set in after fright. Yet they rarely begin suddenly after it. Besides, it is hard to believe that fright can cause irregular movements limited to *one* side of the body. The speculation I hold is, that there is beforehand *local* impaired nutrition, and that the enfeebled parts fail in the general bodily changes which fright brings about. (That there may be local changes before tangible symptoms, is plain to those who have observed cases of acute optic neuritis in physicians' practice.)

It is clear, I think, that fright "produces" chorea most frequently in those who have the "condition for embolism" (see p. 8). A person will not, I think, have hemi-chorea unless the nutrition of convolutions near the corresponding corpus striatum is slightly below par, and he may even then never suffer from it unless there is some sudden extreme derangement of the whole arterial system (and fright is essentially this).

It will be said that children should suffer hemiplegia as well as hemi-chorea, if there is such a liability to embolism. So they do. One child has unilateral palsy; another unilateral chorea; and another unilateral convulsions. Children suffering from any one of these symptoms apart from heart-disease—and thinness after the disease has begun—are usually healthy, and their symptoms have therefore somewhat the character of "accidents." However, there is less frequently rheumatism and heart-disease in association with unilateral palsy in children, and less frequently still with unilateral convulsions. Here again is a difficulty.

[1] See note on Arterial Regions, Med. Times and Gazette, Aug. 15, 1868, p. 177.

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