

ON CHOREA

OR

ST. VITUS'S DANCE

SECOND EDITION

OCTAVIUS STURGES

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Royal College of Physicians  
from the Author.

CHOREA.



# ON CHOREA

OR

ST. VITUS'S DANCE IN CHILDREN.

BY

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*Second Edition, Revised, and partly Re-written.*

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## PREFACE TO SECOND EDITION.

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IN this second edition, besides general revision, the part devoted to symptoms and diagnosis is somewhat amplified ; the chapter on heart symptoms has been re-written, and a short account added referring to chorea in school children. The Tables of the Appendix, occupying much room in the first edition, are now given in abstract only, and in some other ways the matter of the book has been abridged.

Further clinical inquiry, with reference especially to the younger children, has led the author to modify some of his earlier views as to the rheumatic connection of chorea and the significance of its cardiac phenomena. Certain alterations have been made accordingly in the chapters relating to these subjects, and the question of pathogenesis has been reconsidered. Such changes, however, leave undisturbed that account of the affection which refers its main symptoms to functional derangement.

WIMPOLE STREET

*June 21, 1893.*



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## PREFACE.

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THE object of this book is to place before the reader such an account of Chorea, and of the theories which prevail concerning it, as shall enable him to form some judgment as to the nature and suitable treatment of a disorder which is not very common in ordinary practice, yet subject to so many varieties that the study of a few examples gives no adequate conception of its true character.

The first part of the work is occupied with a description of Chorea and of the several hypotheses which have been provided to explain it; there follows an examination of these theories in the light of admitted facts, together with an account of that view of the pathology and treatment of the affection which a full consideration of its phenomena seems to suggest. The illustrative cases given in the last chapter together with the Appendix furnish some of the evidence upon which these views are based.

The purpose throughout has been to show that Chorea is in large measure a disease of function due to preventible causes; that the nature and circumstances of children

render them apt subjects for such disorder; and that, while it is beyond reasonable expectation that the course of childhood should be kept altogether out of its reach, we are not without guidance as to the sort of training and management which affords the best security against its attacks and the surest means for its cure.

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Graphic Representation of Chorea in a Girl aged 12.

I am glad to hear  
of your recovery from  
chorea and hope  
that you will be  
able to write  
from  
Chloris

I now take the  
pleasure in writing  
to you to thank you

This facsimile of handwriting well indicates that precarious and ever-varying control which is the characteristic feature of all movements of the higher muscles in chorea. One word will be nearly illegible while in the next the letters are fairly formed; consecutive letters are not always joined and sometimes they stand far apart, and there are sundry superfluous strokes.

The second specimen was written only a few days after the first and shows but faint traces of infirmity, namely lengthened up-stroke and unequal size of words (see p. 113).

## INTRODUCTION.

### THE PLACE IN NATURE OF FUNCTIONAL NERVOUS DISEASE.

THERE exists in some quarters a disposition to discredit the reference of disease, and especially of nervous disease, to functional causes. Yet if the word disease, itself not easily defined, be left out, functional disorder of some degree, I suppose, everyone must admit, inasmuch as the elements of such disorder are to be met with universally. Granting that there is a wide difference between ordinary emotional spasm and hysteric convulsion ; or between the temporary fidgetiness of a shy child and the enduring agitation of chorea, yet still there is a region intervening which is occupied by muscular disorder of an equivocal kind. The question is, what degree of such disorder is to justify the anatomist in making search for its material equivalent ? No one will answer any degree of it whatever, since that would be to include the whole of mankind, and would involve the absurdity of making disease a universal possession. We are entitled to ask, therefore, how far and in what manner that orderly action of the voluntary muscles which hypothetically represents health may be departed from before disease becomes ' tangible,' if indeed we may not ask yet further upon what authority any particular method of muscular conduct is insisted on rather than another as being its proper pattern, all declensions from which are to be regarded as morbid.

The several conditions of consciousness find a ready and accurate expression in the movements and attitudes of the body. So peremptory, indeed, is this union between mind and muscle that the term 'voluntary' commonly given to the latter is only partially applicable. It is true that the more violent gestures get subdued and toned down by education, and that custom and imitation supply a certain number of conventional and meaningless movements, yet the muscles are not the less expressive on that account. We are seldom deceived as to the nature of emotion by any effort of repression which is used. On the contrary, we can see and measure the force of both.<sup>1</sup>

The monotony and uniformity of civilised life tend of course to reduce motor phenomena to a minimum. After childhood, the full display of passion or emotion is rare, and when it occurs the sympathy of the observer is so far enlisted as to divert attention from the mere muscular condition apart from what it expresses. Thus the several modes of response to as many

<sup>1</sup> Many illustrations could be given of our abject helplessness in this respect. Thus, with all the aid of crowded streets and shop windows, no one ever yet succeeded in appearing unconscious of the neighbourhood of an acquaintance when desirous of passing him without notice; and yet, when in direct intercourse with our fellows we think to pass off sham regard or sympathy, well knowing of ourselves that we should at once detect such counterfeits. Conventional usages of speech and gesture do not really hide the sentiments, but we agree together to pretend that they do. The union between mind and body, whether harmonious or not, is never dissoluble, and it is so intimate that no conflict can be either secret or enduring. The voice of suppressed emotion is really expressing it: the tongue of false congratulation or pretended regard actually stammers. Upon the same principle directness of purpose bears a physical impress which finds instant and universal recognition.

Then my eyes  
Pursued him down the street and far away,  
Among the honest shoulders of the crowd,  
Read rascal in the motions of his back,  
And seoundrel in the supple-sliding knee.

*Tennyson's Sea Dreams.*

different states of consciousness, the spasm of horror, the restlessness of embarrassment, the tremor of fear and sudden disaster, violent and discomposing as they are physically considered, pass without notice, or are noticed only for the meaning they convey, just as the movements of the tongue and lips are not considered in themselves or apart from the words they utter. What would really cause surprise would be the absence of these several emotion movements, or still more, the expression of any one emotion by means of muscular movements such as we are accustomed to associate with another. Thus while many phenomena of movement which are involuntary and disabling are readily accepted as amongst the natural endowments of the individual, no less than are the acts which are voluntary and deliberate, yet each must have its proper motive. Anger excites spasm, fear tremor, mental embarrassment restlessness, and so on. Tremor with no particle of fear to justify it, rigid spasm with no sensation of anger or suspense, a restlessness of the limbs with the mind at ease—these are conditions which require a separate explanation. The ordinary laws of emotion movement do not account for them.

But in thus ascribing to emotion a great variety of muscular phenomena, it is necessary to remember that this particular motive varies widely with the individual. In some temperaments and at some periods of life the most common incidents will suffice to provoke emotion. Familiar illustration of this is seen in childhood, whose ordinary movements indicate a ready and constant yielding of this kind. Again, the spasmodic movements and involuntary gestures of youth are due to such nervous exaltation as is proper to that stage of physical development. And we may say generally that what occasions emotion in one, with its accompanying bodily disablement, will brace and nerve another to his fullest capacity. We

cannot tell beforehand how or when emotion will be excited. We know only that, while different individuals are variously removed from it, there is no one altogether beyond its reach ; and that the response to such emotion is by means of particular movements, spasms, and palsies common to all mankind.

But we need not confine ourselves to emotion alone, but consider as well the ordinary movements of intelligence, those muscular acts by means of which we are always expressing during our waking hours the common current and transitions of thought and sentiment. Such movements, although of the same instigation as the emotional, are so directed and governed by the will as to subserve the purposes of intelligence. The period of life when these movements are the most precise and orderly is that of early middle age, when the muscular frame, while it maintains stillness without obvious restraint, exhibits at the same time by the disposition of the limbs and features muscular alertness along with the ability to use the muscles for intelligent expression without surrendering them to emotional spasm or palsy. He is manifestly the best upon whom this self-imposed control sits the easiest. Rare anywhere, such composure is hardly to be looked for until such time as the comparative calm of middle life has been reached. And what always finds place in such a deportment, side by side with the various sentiments that are animating it, is the continual, watchful presence of self-control, a control of which the highest exercise is seen in the power to maintain a vigorous, capable, unconstrained stillness of the whole frame.

This rare perfection of muscular government, I say, is the end and outcome of a lengthened training having for its object not so much to direct the movements as to restrain them. The limbs which are thus disposed have but lately emerged from their childish condition of impotent movement where they have been swayed about under an influence lower than the

reason yet overpowering it. Even now, should anything occur to disturb the even balance of the mind the restless, motiveless movement of the earlier time would reappear.<sup>1</sup> The best and fullest power of control, indeed, never becomes altogether spontaneous and self-acting. It is not only easily overborne by passion and emotion, but its constant exercise is so difficult and irksome that any occasion for being rid of it is readily taken advantage of. Herein we have one of the uses of laughter, that half-voluntary spasm which in many states of mind affords welcome relief from complete muscular order.<sup>2</sup> Similarly, the best deportment will seek relaxation by means of a variety of distortions, stretchings and grimaces so soon as these may be indulged in without observation. Such acts are proportioned, as everyone will admit, not to the mere bodily fatigue, but rather to the degree of constraint that has been called for. Extreme ceremonial or extravagant gravity needs a larger measure of such relief than the common business of

<sup>1</sup> It will be seen in the sequel that familiar and widely recognised facts such as these are expressed with great variety of phrase. Thus, for example, Professor Bain speaks of a 'central spontaneous energy or activity as a fundamental and permanent property of the system.' 'The active display of the muscles,' he says, 'is most usually and abundantly brought into play by the stimulus of our various feelings, yet is there no reason for supposing that dead stillness would be maintained if outward prompting were withheld.' He further speaks of the restless activity of childhood as 'only to be accounted for by a central fire that needs no stirring from without.'—See Bain on *The Emotions and Will*, part ii., chap. i. What I desire of the reader throughout this discussion is that he will separate facts from expressions, and not accept either metaphor or technical phrase in the light of new knowledge.

<sup>2</sup> It was several times remarked during the Franco-Prussian War that men, after undergoing exceptional danger without bodily injury, would be more than usually alive to jokes, laughing immoderately on slight occasion. Lady Butler, in her famous picture, 'Quatre Bras,' has made use of this observation. There are many similar instances of emotion violently restrained, yet taking advantage of some small opportunity to escape under a disguise.

life. The circumstances may be so ordered as to make the natural craving for this kind of indulgence quite irresistible, insomuch that it has to be yielded to at once, in public as well as in private, and gets noticed and stigmatised accordingly.

And along with this careful control of movement in the presence of others and its welcome relaxation afterwards, we have to consider the obedience that is due to a conventional code of conduct. Although, as we have seen, there can be no actual uniformity of muscular response, but each of us must act after his own prompting, yet is there a strict pattern set up for general imitation. Whoever errs from that pattern, whether by too ready or too violent a display of movement or of spasm, soon finds that these extra acts of his, which would be accepted and allowed if appearing upon proper occasion, subject him to notice, and (during the tutored period of life at least) to rebuke and ridicule. Muscular obedience becomes more difficult when it is thus observed upon and mistrusted, and the loss of public confidence is an extra incentive to error in movement. These new conditions must tell directly on the side of disorder.

It thus appears that muscular response varies very widely with the individual, the time of life, and the circumstances. We can give no stricter account of it than this: that it will be such or such, according as the conditions in their different associations combine to make it. With certain methods of movement and of spasm, ascribed generally to emotion, we cannot say beforehand what is the material which each individual will find sufficient for emotion, the very same cause which disables one bracing up another to his best response. With certain conceptions regarding the voluntary muscles as obedient agents of the will and moving only at its bidding, we find, in fact, that their natural and untrained state is one of constant movement, which is so imperative that the best



discipline, far from controlling it, serves rather to disguise its necessity by giving it some show of purpose. With a certain ideal pattern as to the demeanour which is the fittest, we never actually attain such demeanour; it is not even approached except by long and careful training, and the effort that holds it is always liable to relax. With certain aids to orderly muscular conduct provided by education and public sentiment, it is the common experience that, after a while, the preservation of a set attitude becomes intolerable, while the help which outside observation gives at first to order and stillness, it gives at last to disorder and involuntary movement. I am not asking now how much or how little of muscular disorder actually arises from this wide reach of emotion, or this infirmity of control, or this irksome duty of having to conform to a pattern; I only say that we have here the elements of disorder—material out of which it is inevitable that disorder should spring and grow. To what degree such disorder may spread<sup>1</sup> under the most favouring conditions, or what particular phenomena of so-called disease may be thus accounted for, is a separate question.

And while we thus perceive that the muscular control is variously evaded at the successive periods of life, and take it for a law that the novelty of childhood, the access of new passions in youth, and the monotony of old age should each in turn serve to produce the characteristic movements and attitudes of those periods, it is easy to see besides, how, in a community like ours, the several causes of muscular dis-

<sup>1</sup> A newspaper correspondent at Plevna was an eye witness of the assault on one of the Turkish redoubts, and with the aid of a field glass wrote down the incidents of the fighting as they happened. At a critical juncture his arm became so tremulous from excitement that he could no longer hold the glass. Still watching what went on he had henceforth, to his great loss and annoyance, to trust his unaided sight, so severe and genuine a spasm having possession of his so-called voluntary muscles.

obedience and disorder should press unequally at different ages. It is to be observed, especially, how the motor infirmities of early life get confirmed and intensified by the treatment they meet with. Thus, for example, the natural restlessness of childhood is directly fostered in having to bear the additional strain of observation and rebuke. Unfortunately the prevailing methods of education, though designed to repress movement, serve in fact only to disfigure it. With school children, for instance, the attempt to enforce stillness of the body at the same time that the mental capacity is being tasked, does direct violence to the natural laws of movement.

This extra and artificial strain, which is thus put upon youth in general, operates with its utmost force at certain stages of development and under certain social conditions. There is a time in childhood when the desire for movement is the most active and the least within control; there is also a time when, by our conventional rules of education, the penalties that are put upon movement are the most severe. The nearest coincidence of these two periods (both of which may be fixed approximately), ought to indicate the age when movement disorder is the most frequent and conspicuous. Similarly, at a somewhat later period of life, with the fuller control of movement which age naturally brings, we should expect a comparative calm, until upon reaching puberty a new source of disturbance would have to be encountered.

Such conclusions, I repeat, must be brought to the test of facts. Is the disorder which we thus prefigure actually found; and does it attach by preference to the periods here indicated? Does it prevail more or less according to sex, temperament and social condition, or everywhere alike? Are the particular muscles it affects those that emotion selects, or those affected in structural disease? Is the actual search for its structural basis successful or unsuccessful? The answer to

such questions involves the investigation of the several diseases which, rightly or wrongly, have been classed as functional; and does not concern us now. It is evident, at least, that such an investigation may be undertaken free from any antecedent objection, and that functional disorder, even though it be beyond the field of the microscope, is not unsubstantial in the sense of being unreal.

But the varieties of muscular conduct are not confined to the circumstances of youth, or the special urging of emotion. We have to take into account, besides, such deformities as arise from inherited and acquired tricks and habits, from the natural tendency of human beings to copy one another, and from modifications in the use of the muscles which spring from their education and frequent employment in some particular way. Take any man or woman, out of the crowd, and observe how many are the involuntary acts and spasms which contact with the world has produced or exaggerated; how many movements are superfluous, purposeless and imitative; how complete is the transition, in gesture, and demeanour, and manner of response, when under observation and in solitude, when inspirited and dispirited, anxious or assured. In all these varieties of movement response, which is to be regarded as the normal and healthy one? Is it that in which the muscular response is the quickest and readiest, the state of body always verging on spasm and ready to exhibit emotion by means of over-movement; or is it where the muscles are under full command and no expression is allowed to escape except such as the will and judgment approve, the rarest and least attainable of all human conditions?

The full significance of such considerations as the foregoing will be best seen by observing how the case stands with the most sensitive muscles of all, namely, those of the face. If only the principle be admitted that there is nothing special in

the movements of the face muscles excepting in their alacrity of response, and that a mental impression of whatever kind needs only to be deepened a little to make its motor response spread from the features to the limbs, we shall see then, in the study of this particular region, that spasm, and paralysis, and numerous acts of disobedience on the part of the muscles are part of the daily life of everyone; that, indeed, the precise conduct of the muscles can only be predicted by a knowledge both of the will that informs them and of all the circumstances in which they are placed; that to speak of the normal condition of the muscular system as regards voluntary movement is to speak without precise meaning.

But, while we are thus compelled to admit motor disorder as a necessary ingredient of life, we do not admit it indefinitely. There are the muscular phenomena of functional origin—spasm or tremor, or palsy—and there are the very same conditions due to substantive disease; nor would it be difficult to point out the different associations of the two. We are concerned at present, however, only to show that there is a place in nature for functional nervous disease; our next object will be to make out the claim of a particular affection to occupy that place. There is in the mind of each of us a certain pattern of muscular capacity and muscular control as it ought to be, and there is the larger and less definite pattern which comprehends the actual range of muscular conduct under all circumstances. It is because these patterns are not co-extensive that we need the service of some expression, whether functional disease or any other less offensive to the histologist, to express the difference between them, and occupy the ground which the narrower of the two leaves uncovered.

While, therefore, there is no antecedent objection to this term functional, but, on the contrary, an obvious need and place for it, it is not every disorder of which the material element is

unknown that can rightly be admitted even provisionally into that class. The diagnosis of functional disease, as such, is not always easy, nor is the term itself explicit. Disease is but a name that has been bequeathed to us like many more with no conditions attached that we need at all regard. You may recognize its presence in whatever quickens the heart or flushes the cheek since every such event implies material change, or you may say on the contrary the variability of function is so wide under appropriate stimulus that we cannot identify disease except by its material products. In the one case you hypothesise an individual who is only in health in such mood and circumstances as you arbitrarily determine. In the other you make functional change, apart from disease, of itself a source of suffering and even a cause of death.

The practical question must always be not merely whether the derangement be functional or organic (for we know that the two are necessarily correlated), but under which of these two aspects are the particular phenomena, in their then stage, to be best interpreted. The observation of disease in its functional relations, if less complete than the other, has greater security, for the facts appealed to are exhibited in their life and working without undergoing in the analysis that process of dissection which implies death and may involve mutilation.

## CHAPTER I.

## GENERAL DESCRIPTION.

Pedigree—Definition—Parts affected—Diagnosis—Degrees of severity—Mental involvement—Temperament—Age—Sex—Adult and senile chorea—Geographical distribution—Heredity—Causes—Prognosis—Varieties—Progress—Paralytic symptoms—Recurrence—Connection with heart disorder—Sequelæ.

IN attempting to give some account of the history and relations of chorea, I deem it unnecessary to describe the various dancing epidemics from which it takes its name. Chorea for us begins with its definition by Sydenham. It may, indeed, at first sight seem strange that a movement extravagance of the Middle Ages, having its origin in pious fervour, finding its subjects amongst the emotional and the credulous, and its remedy in some supernatural agency, should have transmitted its name to a purely muscular commotion of children to which the patients are often themselves indifferent, while they are but ill-furnished with that religious sentiment which provided both the cause and the cure of the ancient disorder.

It is remarkable that every epidemic of the kind of which we have any account grew and spread out of material of its own providing. It was never that with the decline of the disorder the terror of it subsided; it was always the reverse of this. The terror died first, and the epidemic could not long survive it. It disappeared as soon as it ceased to inspire the alarm or attract the attention, which was its essential factor. The gesticulations and dancing (although in great measure

involuntary) had the strength taken out of them when this popular tribute to their reality was withdrawn. So long as the priests' exorcism or pilgrimage to shrines was held to be necessary, so long, to the great annoyance of their victims, the muscular contortions flourished and spread; but they could not survive neglect. It will be seen from Hecker's<sup>1</sup> account of these popular seizures that they did not decline from want of subjects, but from a change in the public sentiment. The patients who came latest were discredited. They were not imposters any more than the others, except so far as the knowledge that they would be thus accounted tended to make them so. The same affection, that is to say, neither more nor less real than at first, had lost the necessary incentive of public notice and sympathy.

The current of opinion changes, but men and women remain the same. There is ample evidence in the experience of the present as well as in the dancing and jumping manias of the past, that conscious convulsive seizures need external support for their perfect exhibition. Wanting this, the violence of the seizure is diminished. Thus, while the contagion of other sufferers, and the observation that bystanders are disturbed and impressed, combine to make the circumstances most favourable for the development of such epidemics, the knowledge that the particular symptoms will be recognised only to be discredited, and that those who yield to them are neglected or despised, make the circumstances most favourable for their suppression.<sup>2</sup>

We are not too readily to assume that the chorea we are

<sup>1</sup> 'Epidemics of the Middle Ages' (Sydenham Society).

<sup>2</sup> Upon this subject, in reference not only to popular movement distortions, but also to witchcraft and modern miracles, the reader may be referred to Lecky's *History of Rationalism in Europe* (vol. i. p. 89, et seq.), where it is well shown that the belief in all such phenomena, genuine alike on the part of performer and observer, required for their due exhibition the joint operations of the two. They resulted from 'a general

now to consider is altogether different from these epidemics of old. The very same form of muscular derangement that Hecker describes is with us still, in such dwarfed dimensions as the altered circumstances permit. Its motives and methods of cure are the same now as formerly, and if it is less common or less contagious, the reason is to be found in the wider spread of education. But the healthier public sentiment which has overwhelmed in neglect and contempt, together with witchcraft and sorcery, the various movement extravagances due to ecstasy and fanaticism, has brought into greater prominence the muscular disorder of childhood.

The first mention of chorea indeed as a disease of children coincides with the time when the growing influence of education had acquired strength enough to deprive the popular epidemics of their necessary motive. Along with this change the disorder ceases to be regarded from the old point of view, and descends to a different class of subjects. The notion of demoniacal possession is abandoned, and attention is rather directed to such features of the malady as tend to link it with actual disease. It is due to a *materies morbi* in the blood, to the lodgment of fibrinous clots in the brain, to a new development of articular rheumatism. Such altered attitude of the observer makes an alteration as well in the thing observed. Choreia has changed its shape for reasons which we can partly explain, yet some of the old lineaments are still discernible. The motor disorder which now plagues our children still clings to the same sex as of old, it chooses by preference the same temperament, and is apt to be aroused by precisely the same causes.

predisposition to see Satanic agency in life which grew from and reflected the prevailing modes of religious thought, and declined only when those modes were weakened or destroyed.' With all the distinctive features of our present chorea, we shall find in the sequel that this affection in many of its modes is influenced from without.



Of the many attempts that have been made in our own day to *define chorea*, none can be regarded as altogether successful. It has been described as ‘an irregular convulsive action of the voluntary muscles,’ but it is not convulsive, and not always confined to the voluntary muscles; as ‘a tremulous, irregular, and ludicrous movement,’ but there is nothing of tremor about it, and not much that is ludicrous. Sydenham’s comparison of chorea to the feats of Merry Andrew is familiar to everyone.

The more modern definitions are confessedly faulty and inadequate. In his treatise upon the subject Professor Ziemssen declares that it is only possible as yet ‘to aim at a definition.’ In his own description he is chiefly careful to discriminate between true chorea and the so-called chorea major; the latter as he believes being ‘the product of hysteria or of genuine psychoses and cerebral maladies.’<sup>1</sup> By true chorea he understands ‘a neurosis of which the seat (as it seems) may sometimes be the brain alone, sometimes the entire nervous system; characterised by incessant twitchings or jerks of groups of muscles, which are sometimes spontaneous and sometimes excited by voluntary impulse, which occur almost exclusively in the waking state, and are accompanied by a more or less developed psychical disturbance.’ It may be questioned whether such definition really pictures the affection to anyone not already familiar with it. To ordinary observation, at all events, chorea consists in an exaggerated fidgetiness. It is an extravagant exaltation of that continual unrest which is the natural characteristic of childhood. Its movements, that is to say, resemble the emotional, the same muscles being affected in the same kind of way. Choreia, therefore, may be imitated as emotion may.<sup>2</sup>

<sup>1</sup> Ziemssen’s *Cycl.*, vol. xvi., p. 418.

<sup>2</sup> Choreia, like hysteria, may be seen on the stage far more accurately than in Sydenham’s description of it. Audrey is chorice in conversing with Touchstone. Even in every-day social intercourse there is the

But up to a certain point only ; extreme chorea, like extreme emotion, is beyond imitation, for there belongs to both a degree of agitation which can hardly be assumed, or only partially and for a moment.

Consistently with this comparison the muscles of the *upper part of the body* are much more often affected than the rest, and *the hands suffer most of all*. The thoracic muscles seldom escape altogether, but it is only exceptionally that their concern in the disorder is a prominent feature. The lower limbs are never affected alone, although they may be the first attacked.<sup>1</sup> In the chorea of elder children and of grown-up girls the face is almost always concerned. The more disturbed limbs, or those in which disturbance lasts the longest, become weakened, the loss of power being sometimes very obvious. Certain groups of muscles indeed may appear to be altogether disabled. Thus the hand may hang from the wrist, and for the while be incapable of full extension, or, less commonly, the arm or the leg muscles may become relaxed and useless. Such cases are not less favourable than others. Complete paralysis is rare,<sup>2</sup> and, as a very general rule, there is neither failure of nutrition, loss of tactile sensibility, nor alteration in the muscular response to the induced or continuous current.

movement of the fingers when in animated conversation, fumbling a chain or a locket, or crumbling bread at the dinner table. It is commonly known and taught that a speaker may keep his words within due moderation by resolving beforehand to keep his hands quiet, clasped together, or resting on the desk before him. The mind is so fettered to the body that command of one insures the due order of both. The art of declamation is devised to train limbs which *must* move somehow to move after a graceful manner. But observe the speaker with his hands behind his back and you perceive that the fingers are agitated by a true chorea.

<sup>1</sup> So Romberg. *Sydenham Society's Trans.*, p. 56.

<sup>2</sup> In the paresis of an arm, for example, when the hand, held over the patient's face by the physician, is suddenly released, it rarely happens that the fall is so complete and unbroken as to hit the child's face.

Of diagnosis it is not necessary to say much in this place. Restlessness without spasm, affecting most the upper limbs and seen in its most characteristic form in the hands, with more or less muscular weakness but unimpaired intelligence—that is chorea, be it general or partial. On the other hand, movements that are rhythmical or spasmodic, movements or twitches repeated at equal intervals, movements which, though not violent, are quite beyond control, the restlessness of imbecility—all these are distinguishable from it. It must be added that certain limb movements of late life, though in themselves hardly differing from what we call chorea in children, are yet altogether separate from it in their permanence, in their having often an organic basis, in their hereditary character, and clinical associations.

Diagnosis may be puzzling where the disorder begins elsewhere than in the upper limbs. Thus, marked unevenness of respiration has been taken for paralysis of the diaphragm; inability to speak for commencing brain disease; tumbling about and suddenly falling for some form of paraplegia. There is sometimes much difficulty with young children, in distinguishing the overmovement of idiocy from that of recent nervous disturbance.

Chorea is commonly more marked on one side than on the other and not seldom it affects one hand, or one arm, in especial.<sup>1</sup> For reasons that will presently appear, much stress has been laid upon chorea confined to the arm and leg of one side, so called *hemichorea*. M. Sée, in 154 cases, found chorea confined to the left side, or more marked there, in 97. But this is obviously not to the point, since it fails to distinguish hemichorea in any accurate sense. Dr. Pye Smith, however, found 33 out of 150 *confined* to one side. For myself, I

<sup>1</sup> Compare with the statements here given the figures quoted in the Appendix in reference to the limbs first affected (Tables A and B).

can recall but few cases of chorea of any severity continuing throughout its course limited to one side of the body and including both arm and leg. That chorea commonly begins in one side, all admit; that it may continue confined to a single limb until it gradually disappears, may be admitted also; but a *violent* chorea of one side, in which the other side in no degree shares, is certainly very rare.

The *speech defects of chorea* are of many kinds and degrees. Sometimes speech will be rendered difficult and uncertain owing to imperfect command of the respiration, causing occasional arrest in speech, or the words to be violently jerked out. At other times there is difficulty in setting the vocal apparatus going; the answer to a question will be delayed, as though the other muscles were so busy as to leave no time for speech. In extreme cases of this kind, the lips will feebly move, but the attempt to utter word or sound will go no further. Another kind of speech disorder arises from imperfect muscular co-ordination of tongue or lips, and is seen best in the elder patients, giving to their words that indistinctness and confusion of syllables which is characteristic of drunkards. These speech disorders are in no constant relation to the choreic disturbance elsewhere, and in severe cases speaking may be little or not at all affected. But where speech difficulty occurs, it is commonly an early symptom, and may be the very first. That it may be rightly appreciated, it is well to bear in mind that speech chorea, like chorea in other parts, is a muscular disorder. Aphasia, or any other form of misuse or disuse of speech due to cerebral defect, and not to faulty working of the apparatus of speech, is not met with.

*Defects of deglutition*, less common than those of speech, follow much the same rule. It is not that the power of deglutition fails (although even this may occur), but that in severe cases there is first the difficulty of food reaching the mouth,

and next the difficulty of its being dealt with by the tongue and lips.

Both anæsthesia and hyperæsthesia are sometimes met with (case 17).

The *body temperature*, slightly raised at first, in the more violent attacks; yet, with remarkable uniformity, *tends to fall below the normal*. In the utmost severity of the choreic paroxysms trustworthy observation upon this point is impossible. The highest temperature I have succeeded in taking was 100.6; the lowest, 95.8. Contrary to the opinion of Ziemssen (*loc. cit.* p. 440), that the body temperature is not changed even in severe cases, I believe it to be very rare for the normal heat to be maintained, although the variations are not considerable. The *pulse* is commonly quickened, it is often variable, and sometimes uneven or distinctly irregular. During severe paroxysms it may exceed 120. The state of the circulation, however, will come for separate consideration presently.

Systematic observations on *the state of the urine* in chorea are not numerous, and I have little of my own to say upon this subject. The urea discharge in my experience is very variable, yet not always varying directly with the movements. Dr. Walshe (*Lancet*, 1849, p. 85) was the first to notice high specific gravity, but he did not maintain that this was constant. In a case of acute chorea carefully watched throughout, he recognised four stages in the state of the urine. During the first five days it was febrile, of high specific gravity, deep brownish gold colour, strong urinous odour, and depositing lithates. Secondly came the period with great excess of urea, a symptom referred to the muscular waste due to extra movement. Next, that is on the 26th day and concurrent with marked improvement equivalent to convalescence, oxalates appeared, and lastly an abundant precipitation of phosphates took place, the result of nervous waste.

These observations were in the main confirmed by Todd and Bruce Jones.

*The degree of severity of chorea* may be well measured by observing the patient's posture in bed. One effect of the continual working of the arms and shoulders is to push the patient low down in the bed, thus drawing the head from the pillow, and bringing the body into a horizontal plane. At the same time the child remains constantly on the back, so as to allow full play for the extravagant limb movements. Lying thus, the reckless disposition of the arms in their short intervals of rest is characteristic. They remain, so to speak, where they fall; the flexed arm and wrist lying over the chest, or the limb hanging down loosely over the bed-side.

With its prone position and the depression of the head and shoulders, severe chorea but for its movement would resemble enteric or typhus fever. In the worst cases, indeed, when the violent movement has given place to prostration, this resemblance of posture is complete; and, as in fever, the position in bed from day to day—a little higher or a little lower—is a real indication of the progress to better or worse.

A good working classification of chorea—a rough test of its severity—is found in the ability, or otherwise, of the patient to feed herself. Difficulty in this act has its degrees. Some patients must, for a while, be wholly fed by the nurse. Others, less affected, will contrive to feed themselves a little, and until overtaken by the paresis of fatigue. Much patience and some expertness will be needed in the more severe cases, and it is well to make sure, by measurement, that a sufficient quantity of nourishment is taken daily.

In that form of chorea found amongst young children, which is of long duration but moderate degree—*two varieties* are to be recognised. In the one, the patients are at their worst when conscious of observation, and at their best when

left unnoticed. In the other, there is a considerable power of control when the effort is made; but without it, as when at play or in simple muscular acts, the unsteadiness and disorder become very apparent. This latter variety is by far the more untractable of the two.

Yet, even in severe chorea, it is less true to say that the child cannot be still than that it is unable to maintain stillness. By an effort and for a moment movement can be stopped wholly or partially. And it is curious to notice the effect produced by making appeal to the child's understanding. If, for instance, he be invited *not* to be still, but to move to the utmost, it will commonly happen that the character of the movement is for the time altered, and probably diminished. Similarly, attention directed to a particular limb serves, in a measure, to quiet that particular part; while, at the same time, it will often be noticed that so long as the legs are covered by the bed-clothes and unnoticed, their movement is much less than when exposed for examination.

It is remarkable, and a clue to the pathology of the affection, that in the most riotous agitation of the body and limbs in chorea, *the tongue*, which so often is thus injured in epilepsy, hardly ever gets bitten. The elder patients will sometimes fear such an accident, but they retain will-power enough to avoid it. In like manner involuntary micturition and expulsion of fæces, both common in epilepsy, are here extremely rare, even in the severest cases.

Notwithstanding the inconvenience of chorea, young children take it very composedly. They eat and sleep well, and are in no way mentally disturbed. But it is common to find after the age of ten that at the first onset of the affection, or even before it, the child becomes fretful, capricious, or passionate. With these elder children the temper is continually tried by the disobedience and riot of their limbs. At

a later period still—at and after puberty—the ceaseless agitation, in spite of the will, produces at first a state of mind little short of terror. It is at this time of life particularly that chorea becomes blended, in various degree, with delirium and even mania. (Cases 12, 13, 15, chap. xi.)

It has been asserted that chorea tends in a measure to blunt the mental acuteness ; but in judging of this we are to remember that the instruments of intelligence are themselves impaired, that failure in act tends to produce infirmity and distrust of will, and that the facial deformity often so disguises the natural expression that it is no longer a fair index of the mind. Added to all this, the motor disorder so far occupies the attention that it is difficult and irksome to direct it elsewhere.

Some authors have laid much stress upon the independent variation of two separate elements of chorea, namely, the muscular restlessness and the ataxy. It is pointed out that children with but little restlessness may yet show marked want of muscular consensus, and the converse. It will suffice, in this place, to admit that the fact is so ; its explanation must be sought when we come to consider the pathology of the affection.

As regards *the kind of person* most liable to chorea, independently of the question of age, or sex, or of predisposing diseases, it is enough to say that of those who have written with authority upon the subject some have chosen the dark children, and some the light. All have agreed, however, that susceptible and ‘nervous’ individuals are especially liable, although none believe that chorea is limited to these. The attempt to connect anæmia with chorea I believe to depend on erroneous or too limited observation.

*The favourite age of chorea* is between six and fourteen or fifteen. Its period of dangerous and sometimes fatal severity is from thirteen to about twenty-three. Of Sée’s 191 cases,



151 fell between six and fifteen years of age; only 11 were under six, and 12 over twenty-one. In Dr. Pye Smith's<sup>1</sup> 136 cases, 106 were between six and fifteen. In 71 cases of Dr. Dickinson's, treated at the Hospital for Sick Children, 42 were under ten years.<sup>2</sup> In 177 consecutive cases under my own care, 96 were under that age.

It is thus necessary to amend Sydenham's statement, giving the limits of chorea between ten years old and puberty. It is rather from the period of second dentition to about twelve or thirteen that it prevails, and then suddenly declines. Yet it must be added that the most formidable examples of the affection occur at the time of puberty. If, indeed, such cases were eliminated, the actual mortality of the disease, small as it is, would be very greatly reduced.

Chorea occurs to *girls more than twice, but something less than three times, as often as to boys.* (See Appendix.) This sex difference is the least among the little children and the greatest amongst the elder ones, but it never disappears. The statement that there is no sex disparity under nine years of age is certainly an error. The statistics of many authors upon this point are in substantial agreement. M. Sée, in 531 cases, had 393 females, and 138 males; a proportion of 2.8 to 1. Dr. Hughes has 73 females to 27 males; a proportion of 2.5 to 1. Dr. Dickinson, 50 to 21; a proportion of nearly 2.4 to 1. Dr. Peacock, 66 to 26; or 2.6 to 1. My own tables give 99 to 33—exactly 3 to 1—for the Children's Hospital; and 35 to 10, or 3.5 to 1, for the Westminster Hospital. That there is a somewhat less inequality in the younger children—yet no approach to equality—appears probable from the fact that of 21 children under eight years, of Dr. Dickinson's table, 13 were girls and 8 boys; while of 40 children under the same

<sup>1</sup> See Guy's *Hospital Reports*, 1873.

<sup>2</sup> *Med.-Chir. Trans.*, vol. lix. p. 30.

age of my own, 28 were girls and 12 boys. Of the exact proportion of the sexes at the age of puberty, I cannot speak; but there is undoubtedly a very large preponderance of young women. A severe form of chorea at the approach or completion of puberty in girls, is a not very uncommon occurrence; while the same thing in youths is certainly very rare.

Cases have been reported of chorea occurring to infants at the breast, and even at the hour of birth; but details are wanting, nor is it easy to conceive how the affection rigidly defined can be possible in infancy. The earliest age at which I have met with chorea was in a girl aged two years and eleven months, to be mentioned presently.

The rare and incurable chorea of the aged is, as has been said, distinct from the childish affection we are now considering, being more rhythmical and spasmodic, often hereditary, associated with mental deterioration or actual dementia and having no connection with heart affection.

Professor Charcot, who has called especial attention to senile chorea, has no anatomical pathology for it. He regards it as a merely emotional disease, and quotes examples from the Salpêtrière Asylum to show its origin in distressing events and violent passions, as well as the mental feebleness that goes along with it.<sup>1</sup>

<sup>1</sup> 'On Choreia in Old People.' Lecture by Professor Charcot, *Medical Times and Gazette*, March 9, 1878. See also, in reference to *post-mortem* appearances, 'Cases of Choreic Convulsions in Advanced Age,' by Dr. Macleod.—*Journ. Mental Science*, July 1881. In Chapter XI., case 18, is an example of chorea occurring to a man in middle life, intelligent and unemotional, and with no history of any previous attack.

Dr. George Huntington, of Pomeroy, Ohio (*Medl. and Surgl. Reporter*, Philadelphia, April 13, 1872), described a remarkable form of adult chorea prevalent in the east end of Long Island. Ordinary chorea, he remarks, is very rare there, and in fact hardly known. This adult affection is strictly hereditary and confined to a few families, among whom it is only mentioned with horror as 'that disorder.' It begins in middle or adult life, is commoner with men, occupies years in development, tends to

Along with this adult chorea, mention may here be made of the various muscular tricks and twitches acquired early in life, and which, as the common observation of everyone will admit, cease to be curable when youth has passed. There is hardly an individual of middle life who has not some habit of the kind. These movement infirmities concern chiefly the face, head, and shoulders; and the jerking or twitching of these parts which is often strictly rhythmical differs essentially from chorea. The equality of the intervals appears when the individual is walking, by their always consisting of the same number of steps. There is another form of rhythmic movement which is intimately associated with hysteria and with young women, to which M. Charcot has given the name of 'rhythmic chorea.' In this affection, the arm and leg of one side may be alternately flexed and straightened for a considerable period, or the head may be set shaking, and continue to move with constantly increasing rapidity, to the utter exhaustion of the patient. These and other examples of perverted movement are intimately connected with perverted mind, and belong properly to the subject of hysteria.

As regards *the geographical distribution of chorea*, although we have no statistics to offer, it is known that the prevalence of the disorder varies very greatly in different parts of the world. Over the continent of Europe, though varying much in relative frequency, it is nowhere very uncommon. It is a familiar disorder in France, Germany, Austria, Italy, and European Russia, as well as in the United States. But there insanity, and often ends in suicide. It seldom begins after 40. Its heredity is of this curious form that if it be broken by the failure of one generation to contract it, it is broken for good and all. No treatment avails, and the afflicted families have long ceased to seek advice. The disorder spreads from muscle to muscle, while the mind deteriorates, and the whole man becomes a wreck. Dr. Huntington justly describes this endemic nervous disease 'a medical curiosity.'

are other quarters of the globe where it is far less common. In Northern India I am informed it is very rarely met with, and in Bombay, I learn from a Parsee gentleman, a student of the Grant Medical College, that in the Jamsetjee Hospital for natives, where there are 500 beds, he saw, during three years' attendance, only a single case. Dr. Hughlings Jackson mentions, on the authority of an African medical man, that the affection is very rare among negroes. Dr. Livingstone makes no mention of chorea amongst the diseases he encountered during his travels. It is curious to notice that in countries where extravagant movements and contortions are practised in connection with religious rites, as in Abyssinia, this movement disturbance of childhood, which takes its popular name from a mediæval superstition similarly allied to religion, should be rare. In England the distribution of chorea is very unequal. In some rural districts it is very rare, and while its rule is to favour crowded cities, it is nowhere more common than in the most peopled districts of London.

There is *no evidence to show that chorea as such is hereditary*. In a hundred cases of my own,<sup>1</sup> in which the family history was investigated only 22 (or 21) showed neuroses. They were distributed as follows:—

<i>Parents.</i>					
Father insane in ...	...	...	...	...	1
Mother insane in ...	...	...	..	...	2
„ had had chorea ...	...	...	...	...	2
„ was hysterical in ...	...	...	...	...	1
„ epileptic ...	...	...	...	...	2 (or 1)
„ mentally distressed at time of child's birth					3
„ of very drunken habits at same period					1
					12 (or 11)

<sup>1</sup> 'The Birth and Parentage of Choreia in a hundred Children,' *Lancet*, Sept. 29, 1888

*Brothers or Sisters.*

Had had chorea in	...	...	...	...	3
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*Near Relatives.*

Grandmother insane in	...	...	...	...	1
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Uncle insane in	...	..	...	...	1
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Aunt insane in	...	...	...	...	1
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Aunt had chorea in	...	...	...	...	3
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Cousin had chorea in	...	...	...	...	1
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The rarity of any particular neurosis is here observable and especially of epilepsy. It may be noticed also that only two mothers had chorea of a hundred choreic children—a similar proportion to that of Dr. Osler, who found but seven choreic mothers for 410 children with chorea. What takes from the significance of statistics of this sort is our ignorance of the proportion of insane, epileptic, and drunken relatives we are each entitled to on a fair average. It seems not improbable indeed that any hundred children taken promiscuously would have as many unsound people for their near relatives as those just quoted.

Of the *nervous history and temperament* of children who are the subjects of chorea our information is more precise as depending in large measure on actual observation. Its most striking outcome is the evidence it affords that the nervous disease with which chorea is associated is chorea itself, while epilepsy is conspicuous by its absence.

Of the 100 children reported there were notable nervous antecedents in the following:—

Chorea had occurred before in	...	...	...	29
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Reported as of emotional temperament	...	26
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As showing irritability or noticeable change of temper before the attack in	...	...	...	6
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Headache before attack	...	...	...	8
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Hyperæsthesia of arm in	...	...	...	1
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Again, taking 55 of the elder children (*i.e.*, those over 6 years) in relation to *nervous antecedents* we get :—

Some special nervous disturbance	...	...	25
Naturally irritable, or had become so lately	...	...	17
No known nervous antecedent	...	...	13

How far chorea may be influenced by *social position* is a question of much interest, but also of much difficulty. I believe the fact to be that chorea as a fully developed and recognised disease is very much commoner with the poor, while among the well-to-do we have as many or more examples of *commencing* chorea, which due care and timely relief succeed in arresting at that stage.

*The time of year* when chorea is most prevalent is winter. Spring and summer are about equal, and with autumn begins the increase that continues to the end of winter, or somewhat beyond.<sup>1</sup> The numbers in my own ward at the Hospital for Sick Children from 1876 to 1887 are as follows :—

Spring	...	...	...	37	admitted	with	chorea.
Summer	...	...	...	43	„	„	
Autumn	...	...	...	53	„	„	
Winter	...	...	...	85	„	„	

The only rule that is constant, however, is that which refers to winter frequency, as is shown in three successive years, thus :—

	1884-5.	1885-6.	1886-7.			
Spring	...	7	...	7	...	5
Summer	...	1	...	3	...	5
Autumn	...	8	...	5	...	6
Winter	...	12	...	10	...	9

To sum up then the ascertained facts in regard to chorea,

<sup>1</sup> Sée states ('De la Chorée,' p. 460) that in France autumn and winter show the chief prevalence.

we find its favourite age included between the time of second dentition and that of puberty ; its favourite sex is the female ; the temperament that invites it most is the sensitive and emotional ; it is apt to increase in severity as it decreases in frequency, little children suffering quite commonly, yet hardly ever with a fatal result, whilst of the few instances of adult chorea a notable proportion have died. Further, the disorder is but little if at all hereditary, and more common in highly civilised countries than in other parts of the world. So much is certain ; what is to follow is to some extent involved in controversy. The exciting causes of chorea, its clinical and pathological associations, common sequelæ, modes of progress and of cure, furnish so much material out of which every student of the disorder seeks to obtain a sanction for his own conceptions as to its real nature. I shall endeavour in this place to give the commonly received opinions upon the points just enumerated, in terms which shall be free, as far as possible, from matters of speculation and difference.

Be the exciting cause of chorea what it may, there is commonly a *distinct interval between cause and effect*. It is probable that such interval is really shorter than it appears inasmuch as the disorder is seldom observed at its very commencement. Dr. Hughes<sup>1</sup> made special inquiry into this point in 26 cases. He found that in 13, or exactly half of these, this interval was of not more than a week, in 2 of the 26 it was between one and two weeks, in 3 it exceeded two weeks, and in 8 there was no appreciable interval. Thus in all but 5 the attack followed its presumed cause either immediately or within a week, the latter being the commoner event.

The *immediate exciting causes of chorea* are variously stated by different authors. It is certain and universally admitted that fright and mental disturbance are prominent amongst these,

<sup>1</sup> Guy's Hospital Reports, 1846, p. 380.

causes. Of the 25 children patients of my own quoted above as having been made choreic by some special nervous disturbances (and these are but specimens out of a much larger number that have been under observation) a rough classification would give as particular cause :

- (1) Over-schooling, *i.e.*, school punishment, overwork, the excitement of examination and such like, in 9.
- (2) Definite sources of alarm (*e.g.*, 'locked up in a cupboard,' 'nearly run over,' 'stepped on a snake,' 'chased by men, boys, or dogs,' &c.), the chorea in every instance following immediately, in 16.

It can hardly be doubted that nervous strain is the real exciting cause in a larger proportion of instances than are so chronicled. 'Night terrors,' for example, are so common with children as to have a place in their nosology, yet we seldom learn the precise nature of such terror. And there are many other incidents of child life disturbing enough to their subjects, but habitually escaping our notice.

*Various kinds of pain*, rheumatic and other, headache, articular and other pains, are apt to precede and accompany chorea whether as cause or not. Again, the convalescence of scarlatina, and less obviously of measles, may be disturbed by its appearance. The question of the relationship of acute rheumatism will be discussed in the next chapter. It is beyond dispute that between chorea and endocarditis, whether rheumatic or not, there is during childhood a distinct connection.

That *joint and limb pains* occur not seldom in chorea quite apart from rheumatism there can be no doubt. In the 100 cases referred to above, it was reckoned that 23 out of the 65 non-rheumatic patients had complained of pains and sensations thus :—

Headache	...	...	...	...	...	8
Growing pains	...	...	...	...	...	10
Backache	...	...	...	...	...	2
Cardiac pains, vertigo, local tenderness (each one)						3



It may be safely assumed that this is an under-estimate. Pain that is short-lived and without objective sign is likely to escape record, especially when the sufferer is a child. My own experience includes many instances of patients suffering sudden and acute pain in some one joint, lasting but a few hours and without pyrexia or other symptoms.

There remain a not inconsiderable number of cases whose origin is uncertain. Some of these it is probable have had rheumatism that has escaped notice, and we may safely infer this where there is heart disease or sub-cutaneous nodules. Others again have had nervous disturbance, sleeping or waking, unknown to their parents.<sup>1</sup> The most obscure cases as regards origin are some occurring to young children, and at first mistaken for awkwardness or inattention, an obstinate form of the disorder which gains little by treatment, but recovers at last as it were spontaneously. Strongly contrasted with this is the severe and sometimes fatal chorea of puberty, due often to some obvious mental disturbance, and in the case of girls seldom separable from such cause.

*The progress* of chorea is irregular, and its duration uncertain. In its early development it often shows itself in waywardness, inattention, excitability, change of temper; by such signs, in fact, as come under the notice of mother or teacher, and of which the real significance is not understood. The motor infirmity comes later, and thereupon sympathy or

<sup>1</sup> Pondering these matters in a crowded thoroughfare, I witnessed an incident so apposite to my thoughts and to this subject that I am induced to relate it.

A man in charge of a child in a perambulator was looking in at a shop window when a passing dog put his nose into the child's face, causing it to scream with terror. The man, hearing the cry, looked round to see what had happened, but not until the dog, finding his attentions unwelcome, had disappeared round the corner. Now, supposing this child became choreic, no one save that dog and me could guess at the cause.

alarm takes the place of rebuke. This insidious mode of onset too often subjects the child at the first to undeserved punishment, which, in its turn, tends to aggravate the disorder. Even when overmovement has become apparent, it is habitually misinterpreted. Children tumble about, they have temporary fidgets and fits of temper, and their half-educated muscles are now more now less trustworthy and obedient. No wonder then that the line is sometimes overstepped that separates childish excitement from chorea, without the parents perceiving it, or that they are unable to give any exact account either of the commencement or of duration of the illness.

Recovery is commonly gradual and apt to be interrupted. In some rare cases chorea has suddenly disappeared after mental shock.<sup>1</sup> The rule of the affection is, that after a longer or shorter duration signs of improvement will commence, but that the amendment is subject to interruption and relapse. Slight trouble or excitement, physical pain, the sight of other children suffering similarly, or temporary indisposition short of actual illness, are among the common causes of such relapse. (Cases 6 and 11.) But when recovery has reached a certain point it becomes henceforth rapid and continuous.

The *duration* of chorea is thus too variable to admit of any general statement, and is not proportionate to its initial severity. Dr. Hillier and Drs. Gray and Tuckwell have fixed ten weeks as the average. It is seldom recovered from under six weeks, and, as has been already mentioned, in the case of very young children, it may linger for many months. Duration may be roughly estimated by considering, in a given case, the stage it has already reached, whether waxing or waning. The estimate, however, will be more or less uncertain as the child is more or

<sup>1</sup> An instance is quoted by Dr. Bright. A choreic child on its way to the hospital was frightened in the street; on its arrival the disorder had entirely disappeared.

less timid and emotional, more or less carefully watched and secured from injurious accidents.

Such heart symptoms as are proper to chorea, altered rhythm and mitral murmur, give no guidance whatever to prognosis or to treatment; but when rheumatic heart-disease exists, this of course carries a danger of its own.

Other aids to prognosis are asserted, but they are of doubtful service. It is common to say of many diseases that they improve with the general health; and this is especially insisted on in chorea. It is therein implied both that the health generally suffers in this affection, and that by improving the health you will improve the chorea. Both statements may be doubted. The general health of choreic children is often good, and when it is otherwise it by no means follows that the chorea will get better as the health returns.

It is said further that, inasmuch as chorea comes of insufficient feeding, the prognosis in such cases is favourable because the cause may be easily removed. No doubt this is in a measure true, but it may be questioned whether want of food is ever of itself the cause of chorea, or that supplying that want is necessarily curative. The dependence of the affection on ill-feeding is, I think, exaggerated. Choreic patients are not, as a rule, ill nourished or anæmic, and they certainly do not belong especially to the poorest class.

To these statements concerning progress and duration something must be added in regard to occasional symptoms. One curious incident of chorea, already mentioned, is the intervention of *localised muscular paresis*, a distinct but very variable loss of power in wrist or arm or leg, sometimes of the upper and lower limbs of the same side—seldom amounting to complete paralysis, rapidly recovered from, and entailing no after evil. Appreciable loss of power in one or more limbs is common in the

affection in no constant relationship with the overmovement. In rare instances, the paresis may be so much the more apparent of these two elements as even to obscure diagnosis. Prominence of these symptoms is of no bad augury, and hardly influences the issue.

It is not uncommon for *chorea to recur*, although two, and still more three, recurrences are rare. M. Sée had 37 recurrences among 158 cases. Dr. Dickinson in 71 cases has 18 second attacks, 4 third attacks, and 1 fourth attack. In my own 177 there are 38 which were second attacks, 10 third attacks, 3 fourth attacks, and 1 in which the precise number of attacks was uncertain. (Appendix, tables A and B.)

The *chorea of pregnancy and of puberty* is often of this kind, the same disturbance having occurred in childhood. Dr. Robert Barnes in 66 choreas in pregnancy had 14 such. In a well-known paper upon this subject, this physician<sup>1</sup> has collected from various sources 39 non-fatal and 17 fatal cases of pregnancy in connection with chorea. It is a rare complication, however, happening chiefly to primiparæ, and on the whole not of grave prognosis. It attaches to the earlier months of pregnancy, and often lasts until delivery, abortion or premature birth being not infrequent. Active delirium and acute mania are chiefly to be dreaded, but young pregnant women will get chorea, a revival of that of childhood, of no severity, and with no bad symptoms whatever. (See Cases 14 and 15.)

*Heart affection in connection with chorea* will be considered in a separate chapter. It is universally admitted that a systolic apex murmur is a common symptom in the chorea of childhood. Ziemssen speaks of this murmur as of purely functional origin, due, as he believes, to the action of

<sup>1</sup> *Obstetrical Trans.*, vol. x., p. 147.

See also Lusk's 'Midwifery,' 1888, p. 271.

the papillary muscles. Upon *à priori* grounds he considers that change in the valvular sound 'is more likely in chorea than in any other affection.' He asserts (I believe with perfect accuracy) that there is commonly 'no increase in the second pulmonary sound, no enlargement of the right ventricle, nor any other sign of increased tension demonstrable in the system of the pulmonary artery.'<sup>1</sup> At the same time he recognises 'the residual consequences of an old endocarditis' as sometimes contributing to produce murmur. It is occasionally hard to decide, according to the same authority, whether the murmur be anatomical or purely functional, and he instances anæmia, chlorosis, and acute rheumatism, as exhibiting 'functional disturbances of the mitral quite as obscure as does chorea.' He quotes without adopting the description,<sup>2</sup> the opinion of some writers that 'chorea of the heart consists in irregularity of rhythm audible while the disease is at its height, but neither before nor after.'

The opinion I have been led to form upon this subject, and which I shall seek to justify presently, is as follows:—Accepting the views just stated as to the absence of signs indicating enlargement or change of any kind in the heart cavities, I would insist, too, upon the cardiac symptoms of chorea as being special and unique, a part of the disease itself, coming and going with it, and exhibiting in many instances not only murmur, but marked unevenness of rhythm and even irregularity of action. This cardiac implication is not more frequent in severe than in moderate chorea, and follows no other rule than that it is more common in children than in adults. The

<sup>1</sup> *Loc. cit.* p. 440.

<sup>2</sup> Ziemssen speaks of arhythmia of the heart as 'certainly very rare,' and quotes Romberg's assertion that he could discover no abnormal variation in the movements of the heart in spite of continued observation. Accelerated action of the heart is the utmost he admits. These points will be found discussed in Chap. IV.

irregularity of the heart is in no direct relation to that of the respiration, and seldom gives rise to any subjective symptom. Such description applies to that functional disturbance of the heart to which all young choreic subjects are liable, although many escape it. It must be added that there is at least this connection between chorea and acute rheumatism, namely, that the cardiac disturbance, which at the beginning of an attack of either disease exhibits no more than uneven or irregular rhythm, may develop later a mitral murmur having all the physical characters of endocarditis, upon which, in the event of death, it is often found to depend.

It has been mentioned already that the chorea of puberty is commonly associated with more or less of nervous exaltation. In young women *hysteria is often combined with it* (Case 13). The two affections may occur alternately, or they may be so intimately blended that the one name seems as appropriate as the other. The same sort of sympathy is further illustrated by examples of choreic movement in girls being replaced by tremor or insensibility, or even prolonged immobility.<sup>1</sup> The loss of will control is further shown in a case mentioned by Dr. Handfield Jones, where certain words escaped involuntarily, and to the evident mortification of the patient.<sup>2</sup>

<sup>1</sup> A child (aged six) has lately been under my care recovering from chorea, who exhibited remarkable fixity of attitude, insomuch that she would maintain for a considerable time any posture, however inconvenient, in which she was placed. Thus she would remain quite motionless with both arms extended and the head thrown back; and at ordinary times, although not actually cataleptic, was unnaturally still, even for an adult. Moreover, when set to walk she seemed to want spontaneity, would soon stop, and only after repeated halts and several biddings get round the table. Other similar examples are quoted in the *Medical Times and Gazette*, 'Motor Disorders,' October 19, 1878, and by Dr. H. Jones, 'Functional Nervous Diseases,' p. 362, sq.

<sup>2</sup> The occurrence of painful points, or points painful upon pressure, and of anæsthesia (Case 17), ought to be mentioned in this place. See Ziemssen, *loc. cit.* p. 435.

Among the *rare associations of chorea* is delirium, rising in some instances to such violence as to assume the character of mania (Cases 6, 12, 15). This symptom will sometimes accompany, and sometimes alternate with, or replace chorea, reminding us of a similar conduct on the part of epilepsy. Such nervous implication is found most in the chorea of puberty and of pregnancy, thus adding another distinctive feature to the disorder at this period of life.

It is not uncommon in the violent chorea of adults to find that whenever by a strong exertion of the will the over-movement is violently resisted, the effect of such effort presently appears in the form of emotional agitation. I have conversed with adult patients, who, after their recovery, still retained a vivid impression of the state of mind which they described as inducing them to surrender themselves to over-movement in order to escape emotion. It may be here observed how different is the exercise of volition in chorea and in hysteria, and how marked the contrast between the distress which the elder subjects of chorea manifest towards their disorder and the mental concurrence which attends hysteric convulsion. The muscular movements are in conflict with the will in the one case and in some sort of agreement with them in the other. (Case 13.)

In evidence of the hysteric alliance of chorea, *its imitative tendency* is sometimes quoted. And that the disorder is sometimes spread in this way there can be no doubt. Thus Bucheteau<sup>1</sup> relates that in the course of five days after the admission into hospital of a girl suffering from most intense chorea, eight patients already in the ward contracted the disorder. Dr. Charles West has observed instances of a similar contagion. It must be admitted, however, that this mode of conveyance is rare. In many instances and probably in that

just quoted, it is not so much that the disease is directly imitated as that it spreads by the intervention of terror produced by the sight of it. (Case 12.)

From this aspect of chorea in association with various functional nervous disturbances, we might proceed to consider its connection with structural diseases of the brain and spinal cord. Chorea is sometimes followed by hemiplegia, sometimes it leaves a permanent contraction of the flexor muscles of the wrists, and sometimes such oscillatory movement as is met with in disseminated sclerosis. (Cases 16 and 17). Again, convulsion or tremor has been seen to alternate with chorea, and epilepsy<sup>1</sup> has both preceded and followed it. Of such phenomena we shall speak more particularly presently, it may here suffice to say that they are all exceptional. We are absolutely forbidden therefore from attributing any such meaning to them as they might justly bear were they of common occurrence. What is called 'choreic hemiplegia'<sup>2</sup>

<sup>1</sup> Out of a large number of cases of epilepsy, Dr. Gowers found but twelve who had had chorea, and in only four of those did the fits begin at the time of the chorea. In eight cases epilepsy existed some time or other before the chorea. In the small number of instances where the fits immediately succeeded the disorder, he thinks it probable that 'the impaired nutrition of the motor centres may have left a predisposition to further disturbance.' The exceptional character of the occurrence, however, argues against any such predisposition. See Gower's 'Lectures on Epilepsy,' *Lancet*, vol. i. 1880, p. 355.

<sup>2</sup> In the three cases of choreic hemiplegia related by Dr. Todd ('Nervous Diseases,' p. 312, *et seq.*) and often quoted, the chorea in the first (a boy of nine) was not 'one sided,' nor was the paralysis complete; the patient was nearly well in eight days. The second was a girl of eighteen, who after right-sided chorea appeared uneasy in her right foot, had feeble grasp with that hand, and sensations of numbness in the right arm and shoulder, symptoms not unusual in one of her age and sex. The third case is that of a child of five taken 'with a trembling motion of both arms,' suddenly followed by right hemiplegia, observed only by the mother, lasting only two or three days, and succeeded by right-sided chorea.



is not only very rare, it is seldom complete or enduring. In some alleged instances of the kind, the evidence of antecedent chorea is very defective, while in others it is evident that the preceding affection was not chorea but tremor. A form of paralysis far commoner than hemiplegia is that temporary loss of power on the part of a particular group of muscles which has been just noticed.

Apart from such exceptional instances, what we have to consider now,—what we see ninety times out of a hundred, is the condition I began by describing, a restlessness of the limbs and faces of children easily borne and not affecting the health; involuntary more or less, changeful with the emotions, apt to influence the action and rhythm of the heart; moving the hands and arms in preference to the lower limbs, sometimes limited to a particular set of muscles, but otherwise seldom strictly confined to one side of the body; liable to recur during the period of childhood and youth, while after that time it is often replaced by hysteria, which mixes with it more and more intimately as puberty is approached; an affection of very uncertain duration and uneven progress, yet almost always (in the case of children) recovering completely without entailing any permanent disorder or infirmity.

## CHAPTER II.

## RHEUMATIC CONNECTION.

Analysis of 177 cases—Comparison with other statistics—Conclusions, (1) from general statistics, (2) from comparison of the younger with the elder subjects.

In an Appendix<sup>1</sup> will be found two series of cases bearing upon the question of the relation of rheumatism to chorea. One consists of 132 examples furnished by the Hospital for Sick Children ; the other of 45, furnished by the Westminster Hospital. The 132 cases give 7 who have had rheumatic fever ; 14 to 16 who have had pains, probably rheumatic ; 6 that were doubtful, and 5 where the facts could not be ascertained ; the rheumatic proportion being thus certainly under 34. The 45 cases (the patients being rather older), give 4

<sup>1</sup> In the first edition of this work, the cases here alluded to were set forth at considerable length in tables specifying in the case of each child the age, heart conditions, previous history, and other particulars. In the present work only an abstract is retained (Append., Tables A and B) to represent a large expenditure of time and labour. The reason for this curtailment is not merely the saving of space, but a growing conviction on my part that owing to the nature of child's rheumatism, no record of this sort can be trustworthy. Even our own personal observation does not always enable us to say whether the articular pain of a young child is rheumatic or not ; of what precarious value then must be the account given by the mother as to the nature of joint pain long past ? As is shown in the text, we are now in possession of numerous computations upon this subject from authors of all views, and the aggregate result may be accepted as the nearest approach to the truth that is possible in the circumstances.

who have had rheumatic fever; 5 who have had pains, probably rheumatic; and 11 doubtful; the rheumatic proportion being thus certainly under 20. The want of precision in these figures is betrayed in the large contingent of doubtful cases, but we are probably safe in concluding from them that chorea has nothing to do with rheumatism in from a half to three-fourths of the cases.

As for *the intimacy* of the association, it would appear that of the small proportion having a definite rheumatic history, there are very few showing chorea arising out of rheumatism (or the converse), in such manner as to make the one seem the direct consequence of the other. In about 6 out of the 132, this direct relationship may exist.

Allowing as we must, a considerable margin for error, the conclusion to be drawn from these numbers is not doubtful. Chorea in the majority of cases is apart from rheumatism altogether, and as its direct and immediate consequence, it is a rare event. It may be objected, however, to these tables, as to others, that they are framed on too rigid a definition of rheumatism which excludes some of its proper examples, rheumatic symptoms being easily overlooked in children.<sup>1</sup>

Accepting this observation without reserve, I readily admit that no more than an approximation to the truth is here possible, and that the connection between rheumatism and chorea may be attested by stronger arguments than the numerical. It may be worth while, nevertheless, to compare my own results just quoted with other testimony.

Of continental opinion it may be enough to say that the

<sup>1</sup> Side by side with the statement that the joint implication of rheumatism in childhood may be slight and evanescent, may be put the observations of M. Bouilly upon a febrile condition of children involving the joints called by him 'the fever of growth,' and which, although unconnected with rheumatism, would in all probability be mistaken for it. See Bouilly, *Journal de Médecine et de Chirurgie*, December, 1879.

French and the Germans are much at issue upon this point. Trousseau, as is well known, always insisted upon the intimate union of the two affections, reckoning, however, as evidence of rheumatism any traces of by-gone endocarditis. M. Sée finds chorea and rheumatism coinciding in 61 out of 128 cases; in only 32 of these, however, or one-fourth, were the rheumatic signs decisive. M. Roger has disposed of the question summarily, and in language which renders the labour of calculation superfluous. He believes that 'chorea and rheumatism are one and the same affection under two forms.' On the other hand, Romberg lays no stress upon rheumatism as allied to chorea, and declares his dissent from the views of English writers in this respect; while Steiner, of Prague, records 252 cases, only 4 of which occurred during the course of acute rheumatism.<sup>1</sup>

Statistics upon the subject in the country are abundant. In a well-known paper by Dr. Dickinson (*Med.-Chir. Trans.*, vol. lix.) 71 cases are quoted with as many as 10 doubtful (they are *marked* as 9, but a tenth case is described in the same terms) as to their antecedents. There remain 61, in 42 of whom rheumatism is absent; the remaining 19 had rheumatism somewhere in their history, and in 7 of these it immediately preceded the chorea. Thus 203 cases (these and my own) furnish 139 wholly free from rheumatism, 21 without history, and perhaps 43 who had rheumatism at some period of their lives. In 12 or 13 of these 43 this affection might be regarded as cause, or part cause, of the chorea.

<sup>1</sup> Senator, in his lengthy monograph upon rheumatism, alludes to the risk of heart disease amongst the young, and adds: 'This' (*i.e.*, the heart disease, and not the rheumatism) 'is not unfrequently followed by chorea, especially when the mitral is the affected valve.' This one sentence contains the only allusion to chorea. So little do the historians of rheumatism respond to those of chorea upon this subject.—Ziemssen's *Cycl.*, vol. xvi., p. 57.

These numbers refer to a child's hospital where the utmost age is twelve. General hospitals, having no such age limitation, may be expected to yield a larger proportion of rheumatism, inasmuch as with the larger aggregate of years there is more time for accumulating disease. In two tables of the model medical reports of Dr. Owen, of St. George's Hospital (vol. ix.), will be found a collection of 50 patients, 22 of whom were over twelve years old. Of these 50, 34 were without rheumatism, 3 were too doubtful to be reckoned, and 13 had rheumatism in their histories, in 4 of these last immediately preceding, and presumably causing, the chorea. Adding, therefore, this third list, we get 253 cases, which yield 173 wholly free from rheumatism, 24 doubtful, and 56 who had rheumatism somewhere in their lives, of whom 16 or 17 had the rheumatism in immediate connection with the chorea.

In Dr. Peacock's tables (*St. Thomas's Hospital Reports*, vol. viii.), dealing with 92 cases, only a portion of whom were children, there are as many as 24 or 26 who had had rheumatism at some time or other, the rheumatic symptoms having immediately preceded the chorea in 7; 53 or 54 were known not to have had rheumatism; and the remainder are variously accounted for.<sup>1</sup> The percentage of rheumatism, therefore, is higher in these than in the other tables, yet still the number of the rheumatic is only just over a fourth of the whole.

There remain for consideration the statistics of Dr. Hughes, in the *Guy's Hospital Reports* of 1846 and 1855. It is necessary to observe of these tables that they were not constructed exclusively from cases under the author's own observation, and that, although still much quoted, there is no constant purpose pervading them. In many instances rheu-

<sup>1</sup> From the elaborate character of Dr. Peacock's report I find it difficult to summarise, and would refer the reader to the paper itself, which deals with many other points of interest besides the one here discussed.

matism is not even alluded to. The first series, indeed, is founded upon the supposition, no longer tenable, that the existence of cardiac murmur deposes certainly in favour of past rheumatism. We read, therefore, that while 8 cases only had their origin, 'more or less directly,' in rheumatism, 'there were only 15 (out of the 104 where inquiries were made on the subject) in which the patients were free *both* from cardiac murmur *and* from a previous attack of rheumatism.' Such a statement would not differ very widely from the general experience. Some sort of cardiac disturbance is very common with the younger children. This observation, however, can no longer be accepted as determining the question of the proportion of rheumatism.

In Dr. Hughes' second series of cases, rheumatism finds mention in the notes of but 58 out of 209. The positive fact of its presence is noted 30 times, and the negative fact of its absence 28 times, or about half and half. It is obvious that these numbers fail to represent the actual proportion of rheumatism in the whole body of cases. A small number of examples thus brought together, with a view to illustrate the rheumatic origin of chorea, at a time when such origin is devoutly believed in, will be certain to give undue prominence to that connection. It is only by taking a number of examples consecutively, over a long period, that this disturbing element of choice can be eliminated. Where this is done, the results, as I have shown, are not very various, and non-rheumatic chorea is exhibited in overwhelming excess of the rheumatic, the percentage of the former varying from 75 to 85.

But it is said the rheumatism with which chorea is associated appears in the families rather than in the patients themselves. It is obviously impossible to test this position with any precision. Rheumatism is to be found in every family in the kingdom if we do but seek far enough, nor can

anyone say what is the proper normal amount of it. I have lately noted the point in question in 100 of my own cases,<sup>1</sup> 24 boys, 76 girls, ages from 3 to 12. Inquiry was restricted to the immediate family, parents, brothers, and sisters. The result is :—

Family rheumatism in 25

Personal rheumatism	}	15 near ( <i>i.e.</i> , in point of time).
in ... .. 30		15 remote ,, ,,
Both family and per-	}	4 near ,, ,,
sonal in ... .. 5		1 remote ,, ,,

It will be noted, that of the 35 whose chorea is attributed to personal rheumatism in the above table, only 19 exhibit the disorder in *immediate* connection with that disease. In the remaining 16 rheumatism was remote, and 9 of these had for *immediate* cause distinct nervous shock. On account of these last, clearly attributable to nervous cause, some deduction must be made in the percentage of 35. The rheumatic allowance is still larger than in my earlier inquiries on the same point as given above. It exceeds also by a little, that of the Collective Investigation Committee, which is 26, and largely exceeds that of Dr. Osler, which is only 15 per cent.

It is sometimes urged that the rheumatic element is in part overlooked because in some cases chorea comes first, and rheumatism afterwards. Statistics lend little support to this view. In an analysis of 655 cases of rheumatism by the Collective Investigation Committee,<sup>2</sup> chorea had occurred at some earlier period of life in less than 2 per cent. The claim to kindred which chorea asserts rheumatism does not reciprocate.

By way of a general and impartial review of this subject, Dr.

<sup>1</sup> See *Lancet*, 'Birth and Parentage of Chorea in 100 children,' Sept. 29, 1888.

<sup>2</sup> *Brit. Med. Jour.*, February 25, 1888.

Pye Smith's Table may be quoted as given in his Lumleian Lectures (*Lancet*, April, 1892, p. 954) as follows :—

Chorea was associated with acute rheumatism, according to

Sée ... ..	in	61	of	128	or	48	per cent.
Sturges ... ..	„	20	„	100	„	20	„
Donkin ... ..	„	27	„	104	„	26	„
S. Mackenzie ... ..	„	47	„	172	„	29	„
Herringham ... ..	„	21	„	80	„	26	„
		(or 35 <sup>1</sup> )				(or 45 <sup>1</sup> )	
A. E. Garrod ... ..	„	28	„	80	„	35	„
Collective Invest. Committee ... ..	„	116	„	439	„	26	„
Hughes ... ..	„	106	„	162	„	65	„
Pye Smith ... ..	„	95	„	313	„	30	„

There is reason to believe then from what has been said :

1st. That, excluding the figures of Dr. Hughes, the estimate of observers in this country as to the association of chorea with rheumatism falls between 20 and 30 per cent.

2nd. That the number of instances where rheumatism, in whatever form, is *immediately* connected with chorea is very small; being 4 per cent. in my own cases, 7 in Dr. Dickinson's, and from 7 to 8 in Dr. Peacock's and in Dr. Owen's (St. George's).

3rd. That in the great majority of cases of chorea occurring in children supposed to have been rheumatic, the immediate exciting cause of the chorea is no other than that which suffices to produce the same affection in other children who are not rheumatic.

It is obvious that by no method of argument can these conclusions be made to tell in favour of that *intimate* connection between chorea and rheumatism which some contend for.

<sup>1</sup> The higher numbers of Dr. Herringham are inclusive of examples of joint pains of doubtful character.



Let us say, for example, that 100 instances of chorea yield 25 with a history of antecedent rheumatism. Of these there must be a certain number representing the proper proportion of rheumatic subjects, which we might expect to find in any 100 individuals. What this number should be we know not. Suppose that, on an average, only 5 per cent. of our countrymen get rheumatism in childhood. We have then 20 out of 100 choreic patients charged with rheumatism in excess of the proper number.<sup>1</sup>

The excess, though too small to be adduced in support of any *intimate* alliance, needs to be accounted for nevertheless. The question, in fact, comes to this: Does rheumatism actually confer this extra liability to suffer from chorea at some future period; or is it that the excess of rheumatism is only apparent and due to the fact that chorea shares with rheumatism both heart disturbance and limb pains, so that symptoms which properly belong to the one affection are easily attributed to the other? If the first supposition be adopted it will be necessary to insist (for here the evidence of statistics is decisive)—first, that the rheumatism which predisposes to chorea is seldom connected with it in point of time, a clear interval occurring between the disappearance of the cause and the arrival of the effect; and secondly, that the rheumatic children who become choreic often need the very same immediate incentive to the disorder as do other children—namely, alarm, mental excitement or nervous shock. If the second supposition be adopted, the prevalent belief in the connection of rheumatism and chorea

<sup>1</sup> It is important to add that cases in which chorea arises in the course of rheumatism are so striking and so similar that notwithstanding the comparative rarity of the conjunction it is difficult to resist the conclusion that the one affection actually gives rise to the other. Such examples, although too few to affect perceptibly statistical tables, are yet numerous in the aggregate. And not only so, but the sequence may be reversed, and in the course of chorea, or immediately on recovery from it an attack of acute rheumatism may supervene.

depends upon faulty observation which is sufficiently explained by the circumstances of the two disorders. In the lessening proportion of rheumatism in our later statistics we see the operation of that still recent knowledge which discriminates between the heart affection of chorea and that of rheumatism, while, with the special liability of choreic children to joint and limb pain, we may well accept a certain proportion of them credited with rheumatism without becoming convinced of any real connection.

But the subject is not to be dismissed until the younger children have been compared with the older, for it will appear in the sequel that this asserted pathological affinity finds its chief sanction in early life.

Now in my 177 cases of chorea (namely, 100 just quoted, and 77 analysed in the Appendix), there are but eight under six years old, all but one girls. In one of these history is wanting. Of the seven remaining, three certainly and a fourth probably had had rheumatism: three were exempt. Of Dr. Dickinson's 71 cases, 3 are under six, and in two of these chorea followed immediately after rheumatism. It thus appears that 284 children with chorea include only 11 under six years old. One of these must be neglected owing to defective information. There remain 5 out of 10 who certainly, 6 out of 10 who probably, have had acute rheumatism. With such testimony, the conclusion is inevitable that the rheumatic element of chorea is conspicuous as regards the younger children.

But the numerical statement does not adequately express the strength of this evidence; we have to consider besides the *intimacy* of the relationship in the several instances. The fact that a child suffering chorea, has at some earlier period suffered rheumatism, proves nothing except that the one

disease does not shut out the other, but a single example of the two affections in close union argues strongly in favour of a common pathogenesis. *And such is habitually the nature of the relationship in these young children.* For example, the rheumatic connection of the three youngest of 177 children suffering chorea is as follows :—

1. Annie P., aged  $3\frac{1}{2}$ , 'redness and swelling of the feet just before.'
2. Olive T., aged 4, 'has had rheumatic fever.'
3. Amy G., aged  $4\frac{1}{2}$ , 'rheumatism just before.'

Similarly Dr. Dickinson's 71 cases include one girl (the youngest), under four, who 'had general articular rheumatism just before her chorea;' a boy over five (the next youngest), with 'rheumatism just before.'

If we limit the age to children under five, there are four of such age in my own list, and three of them as stated above are in near connection with rheumatism. Dr. Dickinson has but a single case under five, and it shows the same close relationship.

Since these calculations were made, I have had the good fortune to encounter chorea in a girl a month under three,<sup>1</sup> the earliest recorded so far as I know. Her mother had suffered acute rheumatism a year before its birth. The child had, indeed, no articular rheumatism, but at the heart's apex in the fourth space, half an inch within the left nipple line, a loud musical murmur was audible, conducted into the axilla and heard distinctly behind. If this example may be included with the others, and observation be limited to little children under five, we have, *out of six cases of rheumatism at that exceptionally early age, no less than five intimately blended*

<sup>1</sup> See *Lancet*, Jan. 21, 1888.

with rheumatism; not a large number truly, yet the rare gleanings from a very ample field.<sup>1</sup>

The facts stand thus: if chorea be regarded without reference to age, its connection with rheumatism is neither common nor intimate; but at that early period of childhood, when rheumatism and chorea are rare, when, at all events, they first emerge in recognisable shape, they have a close association. Growth tends continually to weaken this connection, and eventually dissolves it.

What more is to be said on this subject is best postponed until we come to consider the heart symptoms of the affection. The belief in its rheumatic connection is now firmly rooted in this country; we no longer seek to verify the connection, but are rather occupied in constructing theories to explain it. Meanwhile, the two most prominent and unquestionable facts in regard to the causation of chorea are perhaps too little regarded: the fact that alarm or mental strain is its commonest and most direct cause, and the fact that female children are in overwhelming proportion its favourite subjects.

<sup>1</sup> This intimate relationship of the two affections was well seen in the following instance: Mabel J., aged 3 years and a half, under my care at the Hospital for Sick Children (see *Archives of Pediatrics*, May, 1887). The case may be thus epitomised: On admission there was slight choreic movement of right hand and arm, with imperfect control of right leg. A loud systolic murmur with feeble thrill was heard just outside nipple, conducted to axilla. Three weeks after admission, and on the recession of the chorea, acute articular rheumatism set in, and two nodules appeared on left elbow and right olecranon. The physical signs indicated carditis. The child rapidly sank and died. No *post-mortem* examination could be obtained.

## CHAPTER III.

## CHOREA AS A FATAL DISEASE.

Recorded examples of fatal chorea—Age and sex in relation to fatality—  
Exciting causes—Morbidity anatomy—Conclusions.

THE pathology of chorea needs to be discussed upon a wider basis than that which is offered by morbid anatomy; nevertheless, no account of the disorder can be considered complete which fails to take into account the evidence to be derived from fatal cases. The review of a sufficient number of examples of this sort, in addition to the light it may throw on dark places of pathology, cannot fail to be of service in exhibiting the age, sex, and temperament when chorea is most to be feared.

It is well known that a fatal issue is a rare event, so rare indeed that no single observer from his own experience can do more than contribute a very small share of the material necessary for the purpose of drawing any general conclusions. Thus, Dr. Hughes, in so large a hospital as Guy's was able to collect from all the records by various hands, which had been preserved during more than thirty years, only eleven fatal cases. Dr. Dickinson, from a similar record at St. George's, extending over thirty-three years, collected sixteen such cases, while the large field of observation offered by the Hospital for Sick Children during fifteen years furnishes but six. These examples, it must be understood, include those dying *with* chorea as well as those dying of it. Although it is not always easy to distinguish between these two classes, it is certain, as will presently appear, that the cases of death attributable

directly to chorea fall very far short of the numbers just given. It must be added that many of the earlier records are very defective, that there is no uniform plan of tabulation, and that with different observers different points of interest have received mention. On the whole, it may be said that if all the recorded accounts of fatal chorea in this country which are fairly complete and readily accessible were put together, the total number would not be large, and that in a considerable proportion of these the immediate cause of death and the choreic disorder would be found in very remote and uncertain connection. (See Appendix, Table C.)

Such being the facts of the case, I now bring together from various sources 80 cases of death in connection with chorea, or at least with symptoms resembling and called chorea. They are made up as follows:—(a) 3 cases (quoted by Dr. Bright), *Med.-Chir. Trans.*, vol. xxii.; (b) 11 cases (Dr. Hughes), *Guy's Hospital Reports*, 1846 (first series); (c) 7 cases (from same source), 1855 (second series); (d) 34 cases (Dr. Tuckwell), *Bartholomew's Hospital Reports*, vol. v.; (e) 22 cases (Dr. Dickinson) *Med.-Chir. Trans.*, vol. lix.; (f) 3 cases (Dr. Peacock), *St. Thomas's Hospital Reports*, vol. viii.

The points to be examined in these records concern the age, sex, and common exciting cause of fatal chorea, together with its most obvious morbid associations. It will be seen immediately that only a proportion of the 80 cases are given in such detail as to be dealt with in all these respects. Thus, Dr. Tuckwell's account is no more than a short summary occupying one paragraph of his paper just quoted. Dr. Hughes's first series is described by himself as 'very meagre and imperfect'; Dr. Bright's 3 cases, although so often quoted, are not altogether in point; in one there was trismus and tetanic spasm, and in a second paroxysmal attacks described as hysteric. In the several enumerations now to be made, therefore, while 80 is

the extreme number dealt with, smaller numbers will have to serve in regard to particular points owing to defective information. With such deduction the cases available in all respects will not exceed 30, but 30 in the circumstances of this disorder is a large number: it is the sum of fatality out of many thousand examples extending over a long period of years.

Taking in the first instance the question of age, the most striking point of all to those who are in the habit of regarding chorea as a child's affection is *the large proportion of adults and the small proportion of children*. Of the 80, as many as 48 are over thirteen years of age, and the ages of two are not stated. Excluding Dr. Tuckwell's numbers, where such particulars are not given, we have in 46 fatal cases 27 who are over thirteen and only 19 under that age. Of the 27 over thirteen, that is to say beyond childhood, as many as 23 may be said to have died of chorea. Of the 19 who are really children, 10 at the utmost died of it, the youngest of these being a girl of seven (No. 19 in Dr. Dickinson's table, *loc. cit.*). Or, again, excluding Dr. Hughes's first (and defective) series, we should have, out of a total of 35, 20 fatal cases over the age of thirteen, of whom 16 died apparently of chorea; and 15 under thirteen, of whom only 7 died apparently of chorea.

We thus establish the fact, making large allowance for error (which from the nature of the material cannot be wholly excluded), that the mortality of children directly from chorea is exceedingly small, and that seven is the youngest age at which it is known as a fatal disease in a very extensive review. We find also, as we should expect in view of the comparative commonness of chorea in children, that the proportion dying *with* chorea but not *of* it is greater with them than with adults.

Next in regard to sex. In 46 fatal cases (again excluding Dr. Tuckwell's) 34 are females and 12 males. Of the 34

females 8 only were children, the youngest (mentioned above) seven years old, and one described only as 'a child.' As many as 26 were young women, 11 of whom, there is good evidence to show, died of chorea. Of the 12 males 9 were adults and only 3 young boys.<sup>1</sup> These three boys were each eleven years old. It does not appear upon the evidence before us that chorea is ever fatal to males at an earlier age than this, nor have we more than one out of the three whose death is immediately attributable to this disorder.<sup>2</sup>

*The puberty age, then, is the age of fatal chorea.* If we take forty-six as representing the total number of persons dying in this connection, thirty-five would be at or a little above the age of puberty, and twenty-five of these young women.<sup>3</sup> We are thus led to associate fatal chorea with the disturbing incidents of a critical period of life. And the more so inasmuch as in those instances where exciting causes are actually discovered and recorded (but a small proportion, it must be remembered, of the whole number), there is frequent mention of some special excitement of those parts and organs which in the plan of nature are newly active at this particular time. Thus Dr. Hughes, in his second series of cases, finds the genital organs unusually excited in three of the four cases examined, while in Dr. Dickinson's table out of nine dying directly of chorea, and with causes assigned, there are three in whom these causes are distinctly sexual.

<sup>1</sup> These latter are made up as follows: one is in Dr. Hughes's first series (No. 6), another is in Dr. Dickinson's table (No. 14), and died of heart affection 'without return of chorea,' and the third is a case of the late Dr. Fuller's (Case 4, Chap. XI.) that died under my own observation as then Medical Registrar at St. George's Hospital, after five months of almost persistent violent movement.

<sup>2</sup> See Appendix (Table C., Nos. 9 and 10) for two cases in addition to those here reviewed, one a girl, aged 10½, the other a boy, aged 15.

<sup>3</sup> There is one elderly person on the list, a woman aged fifty-four, who had had chorea for four years.—Dr. Dickinson, *loc. cit.*, No. 15.



And not only does fatal chorea attach peculiarly to the emotional sex and the emotional time of life, there are but few instances of it out of the few fully detailed cases which constitute our entire material in this matter where mental excitement does not concur with the bodily over-movement. Such mental disturbance embraces all varieties, from the hysterical to the maniacal. Here again, therefore, a further distinction appears between the fatal chorea of adolescence and the non-fatal chorea of childhood. The latter, even in its greatest violence, is commonly quite apart from emotion. The disorder acquires this mental admixture at the same time that it reaches what may be called its dangerous age.

It will be, perhaps, better in this place to cite instances rather than to quote figures. The condition to which I am alluding is often excited by trivial incidents. Thus, in a case related by Dr. Hughes (first series, *loc. cit.*, p. 390), a man of twenty-five, with violent and ultimately fatal chorea, was 'sensibly hurt and shed tears when the bystanders observed that he could restrain the movement if he pleased.' In a second case, by the same author (second series, p. 250), that of a girl of eighteen, the symptoms 'which were at first slight, were suddenly aggravated by fright caused by a patient in the same ward, and she became almost maniacal.' In a third instance (from the same series, p. 251), a girl of sixteen, where 'there was unusual excitement of the generative organs,' the symptoms at first 'partook of the character of hysteria, the patient appeared strange in her movements, and fancied that others were laughing at her.' In a fourth, a boy of sixteen, 'supposed to be addicted to secret vice, his early movements excited the derision of his comrades. Their conduct depressed him much, and he went suddenly into a fit, after which violent and universal chorea supervened.' In all three of Dr. Peacock's fatal cases mental excitement is a prominent feature.

In one of the very few cases that are recorded of chorea fatal as early as ten (Dr. Dickinson, *loc. cit.*, p. 2), it is related of the child, that a little friend of hers had lately died of chorea, she had watched and imitated the movements, and when she recognised them in her own person made sure that she should meet the same fate. If it be remembered from how small a number of examples these instances are taken, even these few quotations will be allowed to have weight in showing the common habit and exciting causes of fatal chorea.

But while the emotional origin of chorea is thus apparent, as well as the direct influence of mental causes in determining its fatal issue, it is not to be doubted that fatal cases are met with sometimes in immediate connection with acute rheumatism. The evidence upon this point now before us is to the following effect:—Out of thirty-two fatal cases dying *with* chorea, and reported with sufficient fulness to be available (*viz.*, seven of Dr. Hughes', second series, twenty-two of Dr. Dickinson's, and three reported by Dr. Peacock), seven had a rheumatic origin. Selecting out of the thirty-two eighteen who died directly *of* chorea (*viz.*, five of Dr. Hughes', ten of Dr. Dickinson's, and all three of Dr. Peacock's), we get three—a male and two females, *all adults*—where the fatal disorder was of rheumatic origin. These numbers corroborate the conclusion arrived at upon independent grounds in the last chapter (p. 47, 50), namely, that acute rheumatism, although having but a moderate share in chorea, is nevertheless veritably associated with it, and especially with its fatal form at the puberty age.

Coming now to the morbid associations of chorea in so far as they are revealed *post-mortem*, the evidence before us largely bears out the belief expressed by the late Dr. Kirkes in 1863, 'that further experience would still more positively

demonstrate that an affection of the left valves of the heart with the presence of granular vegetations upon them is an almost invariable attendant upon chorea, under whatever circumstances the chorea may be developed.' Subsequent experience has, indeed, shown that the valve affection is by no means invariable and has gone far to negative Dr. Kirkes' theory in respect of it, but the justice of applying the observation to chorea of whatever origin, and not limiting it (as some would do even now) to the rheumatic form, or even to chorea fatal as such, will be seen in what is now to be said.

In all the cases where the heart is described I can find but five where the valves and pericardium are reported healthy, one of these being an elderly woman. Wherever the heart is affected the mitral valve is affected. This condition (to be further described hereafter) occurs equally in those that die in connection with chorea, and in those that die directly of it. Thus, of Dr. Dickinson's twenty-two there are, as I reckon, twelve dying *with*, but not *of*, chorea; of these ten have mitral valve vegetations, and two have not. Of the ten dying *of* chorea seven have vegetations, and three have not. In Dr. Peacock's three cases, all dying *of* chorea, two have vegetations, and one has not. The valve deposit is usually recent, and connected with the mitral valves on their auricular side. (Chap. IV., pp. 61, 70.) The condition has but little apparent connection with the fatal result. It so happens that of the five cases just mentioned as being the only examples of chorea occurring with perfectly healthy heart, all save one, the old woman, are typical instances of death in and by chorea. Three were girls at about the puberty age (Nos. 1, 5, and 8 in Dr. Dickinson's table), and one is a girl of Dr. Peacock's, aged twelve (*St. Thomas's Reports*, vol. viii., p. 29).

Apart from the heart, there is little to be gathered in the material now before us as to the morbid anatomy of fatal

chorea. I shall refer presently to Dr. Dickinson's investigations relating to the minute nervous changes in seven of the cases here mentioned, and to Dr. Tuckwell in reference to embolic plugging. Speaking generally, it must be said that the state of the brain and cord is, in many instances, not described in sufficient detail to be instructive. In a large proportion the brain, in a few the cord, is described as 'congested.' In the case of the boy to whom I have alluded as the single example of the kind in my own experience (Case 4) it was observed that 'the grey matter of the cord was altered and yellow in patches,' possibly sclerosis, but there was no minute examination.

How far the foregoing particulars may serve to fortify or disturb pathological doctrines we need not now inquire. Their practical teaching is obvious. We are justified upon this evidence in looking upon severe chorea at the puberty age, especially in connection with sexual excitement or with acute rheumatism, as a dangerous disorder. It is true that we do not number many fatal cases, but it is also true that we do not meet with many, whether fatal or otherwise. Such nervous exaltation as is commonly altogether absent in the more familiar chorea of childhood, is with these both the sign and the source of danger. They are in a state of extreme sensitiveness, and lay hold of trivial circumstances to aggravate their disquiet. The smallest incident, the conduct of bystanders, undeserved reproach or ridicule, even the sight of some other affected like themselves, will often have its immediate prejudicial influence in a degree which those who are without experience of the disorder may find it difficult to believe. The question cannot but suggest itself, whether in such cases treatment is addressed sufficiently to this mental state.

The prominent fact of our inquiry is the extreme rarity of a fatal issue at that time of life when chorea is commonest,

and to which all our descriptions refer, namely, in childhood. If we exclude the age of puberty, chorea of whatever violence is hardly dangerous to life. Violent chorea is by no means uncommon with boys. Numerous examples of it must be comprehended within the period we are now reviewing. And yet in all that time, and from so many fields of observation, we find but one boy, as young as eleven, dying of the disease after five months' suffering. Choreia, therefore, we may say, very rarely fatal in young girls, is hardly ever fatal in young boys.

The conclusions to be derived from this review are these:—

1. Fatal chorea belongs almost exclusively to puberty, and especially to girls at that age. Its exciting cause in many instances is sexual excitement; but besides sexual causes, mental disturbance of whatever sort has to be reckoned; not fright only, but worry, anxiety, and despondency as well.

2. Acute rheumatism appears as a cause of fatal chorea in but rare instances; yet the occurrence, infrequent as it is, is distinct and unquestionable.

3. Apart from rheumatism, endocarditis of a special form belongs to chorea as such. This condition, however, does not contribute in any way to the fatal issue. It is found equally in those that die *with* and those that die *of* chorea; while in some of the most typical cases the heart has been found absolutely healthy.

4. There is no *post-mortem* condition, except that which concerns the heart, occurring with sufficient frequency of uniformity to be regarded as characteristic of chorea.

## CHAPTER IV.

## HEART SYMPTOMS.

Facts for explanation—The comparative frequency of heart disturbance at different ages—The heart symptoms referred to anæmia and blood state—to sympathy on the part of the heart—to endocarditis—Conclusions.

OBSERVERS are for the most part agreed that a temporary soft systolic apex murmur is apt to arise early in the course of chorea independently of rheumatism or of anæmia; that it is variable in its length, pitch and distinctness, productive of no sensation of its own, and indicated by no general signs of disturbed circulation. It is agreed further that between these cardiac signs and the choreic restlessness there is no correspondence so that the one should increase or decrease with the other, nor is there any particular manner or degree of mis-movement (apart from the question of age) that shows cardiac murmur most or least. These heart signs come and go without notice, and when auscultation has discovered them neither treatment or prognosis is much influenced by the discovery.

There are other characters of heart action as to which there is less agreement. Uneven rhythm (or arhythmia), for example, sometimes but just perceptible, sometimes amounting to obvious irregularity, is a sign questioned by some and neglected by others. But few will deny that the force of systolic impulse is apt to change, and that murmur (if present) varies with it. In addition to these signs the heart's action is commonly quickened and sometimes it is 'tumultuous.' Thus acceleration, uneven rhythm, variable force are the almost

constant characters of the choreic heart in childhood; and to this a varying apex systolic murmur (sometimes loudest at the pulmonary area) is often superadded.

Such symptoms on the part of the heart, in physical characters, limited duration and freedom from injurious consequence, resemble nearly those of so-called functional heart disturbance, yet have they a near relationship to substantive disease. This is shown by the remarkable fact that in the majority of patients dying either in the course of chorea or shortly after it, a fibrinous bead like fringe is found edging the auricular surface of the mitral valve.<sup>1</sup> Coupled with this fact clinical observation clearly proves that the non-fatal cases (that is, the vast majority), though exhibiting such physical signs as have been mentioned during the choreic attack, do not develop heart disease in later life, unless it be in connection with rheumatism. Endocardial inflammation in this connection is both harmless and short lived, and we have to recognise *a form of endocarditis proper to chorea* and separable from the rheumatic in its anatomy, progress and consequences.

In a former edition of this work a statistical inquiry was undertaken with a view to discover the nature and frequency of heart disturbance at different ages and to contrast the younger patients with the older. An abstract of this inquiry (which in its original form was of some length) may here suffice.

The material used for this purpose was the following:—

I. 71 cases (not 70 as stated), quoted by Dr. Dickinson from the Hospital for Sick Children. *Med. Chir. Trans.*, vol. lix.

II. 132 cases under my own care at the chronic branch of the same hospital. (Appendix, Table A).

III. 39 cases (out of 45) at the Westminster Hospital. (Ibid., Table B).

<sup>1</sup> See Appendix, Table C, Cases 9 and 10.

These groups represent respectively :—

I. Young children from 2 to 12 observed at an early period of their disorder.

II. Children of similar age observed at a later period of their disorder.

III. Older children with a few adults.<sup>1</sup>

I. In Dr. Dickinson's table, the number has to be reduced to 69 on account of two cases with ages omitted. In 11 children *under eight years, there is but one free from heart affection, while in 58 of eight years and over, at least 19 are free.* Of 'irregularity,' the 69 cases give 20 examples: 12 of irregularity alone, and 8 with murmurs as well.

II. In the 132 more prolonged cases, 47 have murmur or 'irregularity.' Of 'irregularity' apart from murmur, not including mere unevenness of rhythm, there are but 9 examples, and 6 of these refer to the younger children. These 132 cases therefore, representing the same ages as the foregoing, but referring to a *later stage of chorea, yield a much diminished proportion of heart disturbance, a little over a third as against two-thirds.* At the same time 'irregular' cardiac action largely represented in Dr. Dickinson's table of recent cases, hardly appears in the present list, and is almost confined to the younger children.

III. Of the 39 Westminster Hospital patients (older than the others, and having as many as 15 at or over thirteen years, 2 being young women), only 26 can fairly be reckoned owing to the obvious necessity of excluding rheumatic patients, and those who have previously had chorea. Of these 26, only 7 *have either murmur or 'irregularity,' and only 1 of these 7 is over eleven years of age; while of the 19 whose hearts are unaffected, 15 are over that age.*

<sup>1</sup> A fourth group of 50 cases taken from *St. George's Hospital Reports*, vol. ix., are here omitted as the results are ambiguous, owing to some doubt as to the rheumatic proportion.



It must be observed in reference to these calculations that the term 'irregular' is the only one used to imply arhythmia. It must be taken to signify notable disturbance in the heart's action without taking account of slight unevenness of rhythm. This latter, however, is often the single sign of the heart's implication. According to my own observation, there are very few young children, if there be any, in an early stage of chorea who do not exhibit unevenness and inequality of cardiac rhythm. This will sometimes go on to murmur and sometimes not, murmur which will be almost always variable, apt to come and go, and sometimes finally ceasing before the chorea itself has disappeared.<sup>1</sup>

The conclusions to be drawn from the calculations just quoted are these :—

1. The heart affection of chorea is more marked in early than in later childhood.
2. Observation at a later period of the choreic attack discovers less heart disturbance than at an earlier.
3. Cardiac arhythmia is found chiefly among the younger children.

It may be added, though the point does not occur in the above enumeration, that presystolic murmur and doubling of the second sound at mid-sternum sometimes develop in the course of non-rheumatic chorea, and that those signs like the others eventually disappear.

Now there are two hypotheses of the heart symptoms of chorea, neither of them without difficulty: the one

<sup>1</sup> The question of arhythmia in chorea is denied by some authors, *e.g.*, Walshe and Balfour. Ziemssen says it is 'certainly rare, and hardly anything is published about it,' 'Cyclop.' art. 'Chorea,' p. 436. Bristowe includes irregularity amongst other heart defects. Eustace Smith hardly alludes to it; while on the other hand, Dr. Charles West mentions 'irregularity of rhythm' and 'inequality of force,' p. 209.

asserts that these symptoms indicate a functional disturbance, either in sympathy with that of the voluntary muscles, or else due to an altered state of the blood; the other asserts that they are due to endocarditis, the precise relationship of this inflammation to the motor disorder being variously interpreted. I propose briefly to discuss each of these views by the light of the foregoing observations, and to attempt some judgment between them.

Take first what may be called for short the functional view. 'The apex murmur of chorea,' says Dr. Walshe, 'is plausibly ascribable to disordered action of the muscular apparatus connected with the valve.' In that case the disordered action should extend to the ventricular wall, and to a certain extent it does so. In young children, as has been said, there is often both regurgitation and arrhythmia. But these two signs do not always concur, nor do they vary together. Nor is it young children alone that we have to consider. In the elder patients while murmur is common, any marked irregularity of rhythm is certainly rare. We have thus to suppose that 'the fibres of one and the same muscle are acting regularly as regards the ventricular wall, but irregularly as regards the papillary muscles.' We should be forced to admit, as Dr. Hayden<sup>1</sup> points out, 'that whilst the greater portion of the length of certain muscular fibres contracts with perfect order and regularity, the remaining and smaller portion of the same fibres act spasmodically and out of harmony with the former.' 'No physiologist,' he adds, 'bearing in mind the unity of nerve centres and the community of nerve distribution enjoyed by both portions of the same fibres, would admit such a doctrine.'

There are other objections to this asserted sympathy not less formidable. For if this alleged action of the heart be but

<sup>1</sup> Hayden, p. 274.

an extension of the same disorder that affects the voluntary muscles (an extension, as Dr. Kirkes points out, to which there is no parallel in any other involuntary muscle), then the most violent and general chorea ought to exhibit this cardiac disturbance the most.<sup>1</sup> But it is not so. On the contrary the violent chorea of puberty has seldom either cardiac murmur or arrhythmia, while the chorea of the young child, which is rarely violent and sometimes rather a paresis than a disorder of movement, exhibits both these symptoms very often. And even if these difficulties could be overcome it would still be hard to understand how the great centre of circulation could become the seat of a disturbance at all resembling choreic contortions<sup>2</sup> without giving rise to the most disquieting cardiac sensations on the part of the patient, sensations which so far as young children are concerned are practically unknown.

Not less open to objection are those explanations of systolic murmur in chorea which refer it to muscular fatigue and altered conditions of the blood. This view finds favour with Dr. Hayden. 'The murmur caused by atony and partial yielding of the walls of the left ventricle at the acme of systole have long been associated,' says he, 'with anæmia, purpura, and the excessive use of tobacco.' He is disposed to include

<sup>1</sup> Ziemssen's suggestion as to 'the influence of restless muscular action upon aortic pressure and the working of the heart' (*loc. cit.* p. 437), fails to apply, I am convinced by repeated observation, in explanation either of the heart's irregularity or of its rate. It is, indeed, at once negatived by the admission that the heart disturbance of chorea is in no direct proportion to the violence of the muscular disorder.

<sup>2</sup> 'If such happened,' says Dr. Sansom (p. 334), 'the murmur would present strange variations of character; it would be sometimes soft, sometimes loud, whereas as a matter of fact it presents no considerable variations from hour to hour, day to day, or week to week.' While fully concurring in rejection of this sympathy view of the heart's action, I maintain, at the same time, as stated in the text, that the choreic murmur varies from time to time, sometimes considerably.

chorea in the same category. 'The same relaxation of the muscular tissue,' says Dr. Balfour,<sup>1</sup> 'which is so marked in chlorosis and acute rheumatism is even more exquisitely developed in chorea: hence there is no difficulty in referring the continuous mitral murmur in chorea to the same cause which has been already shown to be capable of producing it in the diseases referred to.'

Other authorities might be quoted to like effect. 'The muscular tissue of the heart,' says Prof. Immermann,<sup>2</sup> 'owing to the altered state of the blood is easily fatigued, and this liability to premature fatigue extends to the papillary muscles connected with the auriculo-ventricular valves. After an undue exertion of the cardiac muscle a temporary paresis of the musculi papillares ensues. In consequence of this the valve flaps intrude into the auricles with every ventricular contraction, that is, a transient functional insufficiency of the tricuspid and mitral valves is established.'

Since this was written, the precise mechanism of the dynamic murmur has been fully investigated by Professors Roy and Adami, as well as by Hesse of Leipsig.<sup>3</sup> It is now known that systolic murmur is produced by the joint agency of valve and ventricular wall, though it is not always easy to account for the ventricular defect in individual cases. The authors just quoted would refer it to altered blood states, anæmia, purpura, and the like. But, suppose we accept the so-called hæmic murmur as due to that cause, what have anæmia or purpura to do with chorea? Dr. Balfour, in the passage just quoted, seems to imply that the evidence of blood change is even more marked in chorea than in anæmia. There is no possible reply to such an assertion, except direct denial and

<sup>1</sup> Balfour's 'Diseases of the Heart,' 2nd Edition, 181.

<sup>2</sup> Ziemssen's 'Cyclop.,' vol. xxi., p. 399

<sup>3</sup> See MacAlister, *Brit. Med. Journ.*, Oct. 28, 1882.

the counter-statement that the blood in chorea shows no sign of change whatever.

But even were it the fact that all choreic children were anæmic, this theory of cardiac paresis from muscular fatigue would still be inapplicable. For it is not, as I have shown, where the movement is most violent and fatigue the greatest that these cardiac signs appear: it is almost the reverse of this. Nor is it, as upon the hypothesis it should be, that after a certain endurance of chorea the heart gets tired and exhibits its fatigue in the way stated. On the contrary, systolic murmur in young children will often appear quite at the beginning of an attack, nor does it make any difference whether such attack be slight or severe.

It would seem then that neither of the theories just reviewed—neither the cardiac sympathy hypothesis, nor the dynamic hypothesis—can be accepted without doing violence, not only to the facts of the case but to well-recognised physiological laws. If the systolic apex murmur of chorea betokens a sympathy on the part of the heart with the disorder of the voluntary muscles, it ought to appear when the disorder is the most generalized and to increase as it increases; if, on the other hand, such murmur betokens a fatigue paresis, it should appear when the chorea is most violent and most prolonged. But it does neither.<sup>1</sup> The asserted sympathy of the heart goes not with the degree of chorea, but with the age of the patient; while the systolic murmur audible at an early stage of the disorder will cease at a later. Moreover, we should be taking but a partial view were we to limit the heart symptoms to systolic murmur. There is besides, especially in

<sup>1</sup> In rare instances, however (as in Case 11), the occurrence of muscular paresis along with a distinct mitral murmur, and the disappearance of the two symptoms at the same time, is an undoubted fact, however it may be explained.

early childhood, the arrhythmia and, most constant of all, there is the accelerated pace of the heart.

The objections I am now urging refer to clinical phenomena. But the anatomical side of the question is not to be forgotten. Were the murmur of anæmia and of chorea ever so similar, there is certainly no similarity between the anæmic and the choreic heart. Characteristic changes may be found in both, but with no sort of resemblance ; and in urging the acceptance of either of the doctrines just mentioned morbid anatomy must be left out of sight.

Let us turn then to that view of the heart symptoms of chorea which refers them to endocarditis. Now, in some instances, as we have already seen, this is not a view merely, it is a fact. The physical signs suggestive of endocarditis have in many fatal cases, as was shown in the last chapter, been traced to their cause in actual valve inflammation. But obviously far more than this is needed before admitting that the heart signs commonly met with in ordinary recovering chorea, and which are seldom permanent or injurious, are always due to a like cause. The discovery of endocarditis in a proportion of cases out of the extremely small number that end in death (Append. Table C, Cases 9 and 10), goes no further than to suggest the possibility that heart disturbance in chorea is commonly so produced. Apart from *post-mortem* evidence, no one would venture to maintain upon the mere showing of certain physical signs, notoriously equivocal, that an affection like chorea having all the characters of a functional disorder, was in reality closely connected with carditis. Furnished with such evidence, however, the case is altered, and particularly when we consider that endocarditis, however rarely revealed, is the one tangible anatomical feature of the affection.

The distinction between organic and functional heart murmurs is admitted to be difficult. Rules founded on hard and

fast lines as to their transmission are of doubtful validity. 'There are few diseases,' says Rosenstein,<sup>1</sup> 'the presence of which is diagnosticated so arbitrarily as that of original acute and subacute endocarditis.' 'A well pronounced functional murmur' says an excellent observer,<sup>2</sup> 'may be as diffused and transmitted as an organic.' We have, indeed, in many instances, just as much evidence, so far as physical signs are concerned, of endocarditis in the course of chorea as we have (and are in the habit of accepting in that sense) in the course of acute rheumatism. In both cases alike a soft and ill-conducted murmur may be proved in the event of death to depend upon valve deposit; and in both alike (I do not say to a like extent) when death does not so intervene the murmur has been known to disappear.

Considerations such as these, with the manifest failure of other hypotheses, compel the comparison of the heart phenomena in chorea and in rheumatism.

Now in the course of acute rheumatism, as in the course of chorea, the heart's action will become tumultuous, a slight bruit will be next heard, and this will presently become louder and be indistinctly conducted into the axilla. After a while these signs begin to recede, and at the end of the rheumatic attack they may be barely audible. What has happened? It may be in the case of rheumatism (but not in the case of non-rheumatic chorea) that pericardial friction has been heard, whereupon the existence of endocarditis is inferred. But pericarditis is an event of rheumatism, not of chorea. And in virtue of this difference the latter wants that interpretation of its physical signs which the former enjoys. Nevertheless the early physical signs of chorea are precisely similar to the early physical signs of rheumatic endocarditis. In both cases they suggest the

<sup>1</sup> Ziemssen's 'Cyclop.,' vol. vi., Art. 'Endocarditis.'

<sup>2</sup> Dr. Nixon, *Dublin Journal of Medical Sci.*, vol. v., p. 575.

oncoming of that inflammation, although, as Dr. Sibson has pointed out, such symptoms of heart excitement are sometimes only the 'threatenings' of actual inflammatory deposit.<sup>1</sup>

Observe that this early likeness, or rather identity of physical signs in non-rheumatic chorea and in rheumatic endocarditis refers to the early stage only. There is no likeness whatever in the later stages. With the occurrence of mitral endocarditis in chorea after the fashion already described, the part which the heart plays in that affection comes to an end. Those later changes leading to valve distortion and consecutive alteration in the heart's chambers which belong to rheumatic endocarditis, have no place at all in that which belongs to chorea. Hence it happens that it is only in the very rare event of rheumatic endocarditis proving fatal at a very early stage that it presents *post-mortem* any likeness whatever to that form of mitral endocarditis—a fringe of tiny beads edging the mitral valve—which is the beginning and the end of heart change so far as chorea is concerned.

The case of chorea, when associated with rheumatism, is not now in question. All will agree that the endocarditis of

<sup>1</sup> Referring to Sibson's expression regarding the physical signs of 'threatened' endocarditis, it is to the purpose to notice M. Hayem's observations on the systolic apex murmurs developed in the course of some fevers and due to myocarditis, as quoted by Dr. Sansom (p. 335). 'Although at first only of slight pronounciation, it may soon become louder and make one believe in the existence of endocarditis. Moreover, it may vary in intensity from day to day or become modified by a change of position of the patient.' 'This murmur,' as Dr. Sansom adds, 'may be accompanied by a doubling of heart sounds and disturbances of cardiac rhythm. All these signs eventually disappear. Now these are precisely the phenomena of the disturbed choreic heart, and 'the conjecture is whether the early, and in many cases the only affection of the heart in chorea, as in the acute fevers, is a myocarditis whereby the muscular fibres become swollen and so far changed that the mitral curtains are imperfectly opposed in systole and regurgitation in some degree may result.' (See Sansom, *loc. cit.*, p. 335).



choreic children who are known to be rheumatic is probably itself rheumatic, and the precursor of further valve change. The endocarditis that is proper to chorea, apart from rheumatism, is of a different sort. It is special alike in its morbid anatomy, clinical course, and sequel. It leads to no disturbance of circulation whatever, is temporary and innocuous, and on that account only by rare accident demonstrable on the *post-mortem* table. But, as was shown in the last chapter, we are not without examples of it, and these exhibit a form of endocarditis which is very rare in any other association.<sup>1</sup>

I have lately collected a hundred cases of valvular heart disease in children examined *post-mortem* at the Hospital for Sick Children during eleven years.<sup>2</sup> The result is as follows:—Endocarditis occurring by itself was found to be very rare. Of *recent* endocarditis, indeed, there was no single example amongst the rheumatic (54 in number), without participation in some degree on the part of the pericardium. Amongst the non-rheumatic (46), there was a solitary example of *recent* endocarditis of the mitral valve alone with no pericarditis or excess of pericardial fluid. This was the sole instance amongst the hundred wherein recent endocarditis was thus confined to

<sup>1</sup> I may allude here to Dr. Sansom's contribution to this subject. 'Papilliform endocarditis,' he says ('Diagnosis of Diseases of the Heart,' p. 332), 'may be manifested apart from any cause associated with rheumatism, and is probably due to a sudden disturbance of the intra-cardiac pressure whereby violence is done to the delicate endocardium. The lesions do not resemble those of rheumatic endocarditis. There is hyperplasia of the substance of the valve itself causing small warty outgrowths. These are only found *post-mortem*, when death has taken place from other causes.' Dr. Sansom goes on to describe later appearances where the outgrowths are 'fibrous and hard,' a condition certainly not met with in chorea. But what I would point out is that this thoughtful and accurate writer sides neither with the anæmic nor with the rheumatic view, but concludes that sudden emotion may be the initiating cause of the endocarditis of chorea (p. 48), a theory to be considered presently.

<sup>2</sup> 'The Rheumatic Carditis of Childhood,' 1893.

the mitral valve, and the child who was the subject of this condition died directly of chorea.<sup>1</sup>

Similarly it has been shewn from records derived from various sources that mitral endocarditis indicated by soft papules or beading on the auricular aspect, is the characteristic feature in fatal cases, and a condition that belongs to no other affection whatever. (Appendix, Table C.)

Thus endocarditis attending chorea is of two distinct forms; in the one, the non-rheumatic and intrinsic, it is only revealed *post-mortem*, usually affects the mitral valve alone, is neither permanent nor injurious to the heart, and belongs to chorea as such; in the other, the rheumatic, cardiac symptoms are soon manifested, permanent valve changes ensue, and these, independently of the chorea, lead eventually to failure of compensation on the part of the heart.

In witness to the fact that the endocarditis of chorea is *sui generis*, that it has no objective signs, and that in fatal cases the manner of dying is precisely the same with it as without it; let me refer to two cases mentioned in the last chapter (p. 57), one reported by Dr. Peacock, the other by myself. In neither were any heart symptoms observed during life. Both died exhausted from chorea. In the one, the heart and pericardium were 'quite healthy'; in the other, 'the inner edge of the mitral valve showed a line of soft beads easily detached.' But for *post-mortem* inspection, this true chorea endocarditis would never have been discovered.

<sup>1</sup> The following is from the *post-mortem* notes of the case:—Muscular substance of the heart normal. Consistence perhaps a little soft. No pericarditis or excess of fluid. The free edges of the mitral valve show signs of recent endocarditis, a thin red line everywhere along edge, with small, low, firm and adherent vegetations which can only be separated with difficulty. Endocardium generally quite healthy. The heart cavities contain a very little non-adherent clot and some fluid blood. ['*Post-mortem and Case Book*,' vol. x., p. 179.]

It is not so that rheumatic endocarditis behaves. In those striking, yet not common, cases where chorea and acute rheumatism occur in close proximity, alternating the one with the other, the morbid changes, products not of the chorea but of the rheumatism, tend sooner or later to mitral narrowing and consecutive changes in the heart's shape and size. In such circumstances the appearance of chorea is of pathological interest rather than practical importance. The heart disease will progress faster or slower in proportion to the frequency of the rheumatic attacks. Those of chorea will not affect its progress at all. That the occurrence of endocarditis, both in chorea and in rheumatism, is some evidence of affinity between the two affections need not be denied, but it happens in chorea in one way, in rheumatism in another way. To say that in chorea endocarditis is a manifestation of rheumatism is no more warrantable than to say that in rheumatism it is a manifestation of chorea.

Some allusion may be made in this place to the views of Dr. Sansom. Admitting, as he does, the fact of an endocarditis in chorea, which is certainly not rheumatic, this physician would explain the occurrence by reference to the sudden emotion, which in a certain proportion of the subjects is the exciting cause of their malady. After combating the opinion that valvular disease in children is always rheumatic, he goes on to describe the physical effects of sudden terror: 'The face turns pale, the heart's action becomes arrested, the arterioles contract. Then ensues a reaction, the heart palpitates violently, and the mechanism of the valves, still for a moment under the heightened blood pressure of the contracted arteries is set into tumultuous action. It is not suggested that the valves are subjected to overstrain during the continuance of the choreic convulsions, but at the time of the immediate phenomena of the fright which is its disposing cause.' 'The endocardial

change is the direct result of violence ; the conditions of blood pressure within the heart cavities must be profoundly altered. In the delicate structure of the endocardium of the valves of the child, this influence is likely to be more pronounced than in the adult. Minute hæmorrhages in the endocardium result in the formation of small papilliform outgrowths ; so the limited papular form of endocarditis is initiated.'<sup>1</sup>

To a theory so ingenious and so justly emphasizing the nervous and psychical origin of the affection, no less than the anatomical fact that the endocarditis of chorea is not rheumatic, it is no grateful task to raise objection. But it will occur to the reader that terror is a very common incident in child life, while it is only a small proportion of frightened children that develop chorea. No heart symptoms are known to ensue from fright in the numerous cases where chorea does not follow. Yet, according to Dr. Sansom's statement, it is the fright and not the chorea that initiates the heart change. And besides, if this theory were valid, the heart symptoms of chorea (putting rheumatism on one side) ought to attach to those children whose disorder has been occasioned by terror and not to the rest. But there is no such distinction in fact. The agency of terror in the production of chorea I should be the last to deny. But there are other agencies not less obvious—overwork at school for example, putting children to task by lessons above their capacity, especially sums in arithmetic. Such patients hardly furnish the examples that Dr. Sansom seeks. There is no actual terror involved in the unsuccessful attempt to work a sum in long division, no sudden pallor of the face and arrested heart's action, only weariness, and at last it may be despair.

It must be remembered also that the most striking illustration of sudden emotion is that seen in young children in what

<sup>1</sup> Sansom *loc. cit.*, pp. 48, 333.

is known in nursery phrase as 'a passion'; they tremble and get pale, exhibit the utmost violence, sometimes ending in slight convulsion, and exactly fit Dr. Sansom's description. But they do not get, so far as is known, minute hæmorrhages in the endocardium. It would be surprising to learn that an emotion so common with little children as fright or passion might be the direct exciting cause of valve changes.

The further discussion of this subject will be best reserved for the chapter on pathology. Enough has been said to show its difficulty, and that some violence has been done to the natural character of chorea in attempts to solve it. In so intricate a matter we must proceed cautiously from what is known to what is hypothetical, with the greatest assurance when depending immediately on ascertained facts and the least when within the region of conjecture. In the failure of other explanations we are bound to lay stress on the cardinal fact of the disease, namely, the very common occurrence of endocarditis, taking note at the same time that this endocarditis is *sui generis*, and not to be confounded with the rheumatic. Now without entering as yet into the question of pathogenesis, we have here evidence of cardiac sympathy both with chorea and with rheumatism, and it is expressed in both cases, not only by physical signs, but by the material products of inflammation. But the likeness between rheumatism and chorea which this common sympathy indicates is a likeness merely, in some respects a superficial likeness. If rheumatic carditis, as a whole, be compared with choreic carditis, as a whole, there is more of difference than of resemblance. They begin the same, but they end differently. The rôle of cardiac inflammation in rheumatism is more extensive and more prolonged than in chorea. It is not limited to the endocardium; it goes on to the production of vegetations, and sometimes to ul-

ceration ; and it variously deforms the heart. In none of these respects does choreic endocarditis go with it. This latter, indeed, has no further stage than the bead-like out-growth already described, which now and again the rare accident of death discloses, but which probably in the ordinary course is completely recovered from. The close kindred, or indeed, as some contend, even the practical identity of chorea and rheumatism, is plausibly affirmed by the presence of endocarditis in both ; but the real separateness of the two inflammations is amply shown in the special characters of each.

Nevertheless, it remains true that the heart after its manner sympathises with chorea as well as with rheumatism, and although it is true also that the precise expression of such sympathy is different in the two cases, I cannot but see in the fact of it a solid reason for admitting some real affinity between the two affections. It is, I submit, in virtue of such affinity that we can the most easily, and without distortion of clinical facts explain the remarkable phenomena to which this chapter is devoted.

The conclusions on this subject may be expressed as follows :—

1. The heart affection of chorea is more marked in early than in late childhood, and early in the course of an attack rather than late.

2. Papilliform endocarditis affecting the mitral valve, a condition which, occurring alone, is rarely seen in any other association, is the cardinal anatomical feature in chorea wholly apart from rheumatic endocarditis. Yet it is not constantly met with, is without subjective symptoms, not persistent or injurious, and leads to no consecutive changes of hypertrophy or dilatation. Some of the most striking examples of death by chorea exhibit no cardiac lesion whatever.

3. Thus chorea and rheumatism are both liable to cardiac inflammation, each after its own manner, although the distinction between them is sometimes obscured, owing to the concurrence of the two diseases in the same subject.

4. The heart symptoms of chorea seem best explained upon the hypothesis of some pathological kindred between it and rheumatism.

## CHAPTER V.

## CHOREA AT SCHOOL.

Examples—Mode of origin and development—Points for observation.

IN this chapter, at the risk of repetition, I would set down some simple rules for making early discovery of chorea and roughly estimating its severity. The question of early diagnosis in fact concerns school teachers rather than medical men; for it is at school that the affection begins, it is there that it passes from a mental to a motor disorder, and (whether from blunt perception on the part of school managers, or the want of necessary guidance on our part) often remains unrecognised until it has reached an advanced stage.

An analysis of 25 children under my own observation who exhibited movement disturbance due to some particular nervous overstrain will help to show in what degree school worry is directly concerned. In 16, the causes are definite sources of alarm for which school cannot be held responsible, *e.g.*, being 'nearly run over,' 'chased' by men, women, boys, or dogs (several under such headings), 'locked up in a cupboard,' witnessing some alarming incident, quarrels, drunken violence, &c.

The remaining 9 of the 25 (or over a third) were directly connected with school: *e.g.*, overwork, punishment, preparation for examination, and so forth.

With a view to bringing home to those who are unfamiliar with the subject the precise manner in which school children are thus injured, together with the important fact (which some may be disposed to dispute) that children are often suffered to



remain at school notwithstanding their movement infirmity, I will recall the circumstances of six months' (November, 1890, to May, 1891) hospital experience in these respects.<sup>1</sup>

During this time 40 examples of St. Vitus' Dance were admitted to the Hospital for Sick Children under my colleagues (Drs. Cheadle and Barlow) and myself. I quote the following, all but one are girls:—

1. *Florence P.*, age 11 years 9 months. (February 12 to March 2.)

A second attack (the first attributed to being 'caned and knocked about at school'). She has been worried over lessons, and observed to sleep badly. It was first noticed that she would drop things, and was very restless both day and night. The restless nights preceded the movements by some weeks. On February 1st, the parents noticed in addition that the child had difficulty in writing, and *she was removed from school four days later.*

This girl showed St. Vitus' Dance of moderate severity; quickly recovering on getting rest.

2. *George H.*, aged 10 (the only boy of the eight). (January 23rd to March 26.)

As far back as November observed to be restless, *but not removed from school.* At Christmas much frightened at seeing a woman knocked down and her head cut open. Was much more restless the following day, screaming and crying out at night. *Was still kept at school* notwithstanding, and only last week was punished for inattention at lessons. This boy, though not very bad on admission, became violently agitated sometime later after a dream, weeping and throwing himself about in paroxysms, quite beyond self-control, and fed with great difficulty. He was more than two months in hospital, but eventually made good recovery.

3. *Bridget M.*, aged 11 years and 7 months. (9th December, 1890, to 1st January, 1891.) (Dr. Barlow's case.)

For three months this child has been working for examination, and for two months has stayed in school until eight o'clock, doing home work as well. Lately has complained of headache. Worried herself especially about sums, fearing that she would not pass in arithmetic. The disorder increased to the degree of St. Vitus' Dance on the first

<sup>1</sup> This particular time was chosen, as it preceded the meeting of the Congress on Hygiene, in 1891, at which a paper was read by the author, of which this is the substance.

day of the examination. It thus happened, that as the child sat writing her paper, the restless left elbow by misadventure joggled the writing hand of a boy sitting next her, and helped to spoil his work. She was much upset by this accident, and the movements increased. On admission, the limb movements were not considerable, but the child was unable to walk, and had difficulty in bringing out words. She recovered well.

4. *Gertrude MacS.*, aged 11. (April 28th to May 21st.) (Dr. Barlow's case.)

This was the second attack, and had begun three weeks before admission after she had been in for examination. The disorderly movements were first noticed during writing; they afterwards became general.

5. *Rosa W.*, aged 9 years and 9 months. (March 19th to April 23rd.)

General restlessness and difficulty of speech, not due to schooling in the same sense as the preceding, but to the excitement produced by taking part in a school concert. (Dr. Barlow.)

6. *Kate P.*, aged 10 years and 2 months. (February 27th to April 2nd.)

Has been working hard for June examination. 'Hardly sleeps at all.' The movements have been noticed a month, beginning in face, twitching of eyelids and mouth. *For the first week, however, she was kept at school.* On admission the movements were severe, and the girl could not feed herself. The ease, it will be noticed, was of some endurance, but the child recovered perfectly.

7. *Edith C.*, aged 9 years and 5 months. (November 7th to December 11th.) (Dr. Cheadle's case.)

Alleged cause 'much worried with lessons at night, and finds difficulty in preparing them.' Attention was first drawn to the child's unsteadiness of grasp a fortnight ago. Her walk was next observed to be irregular, and the corners of the mouth twitched. (The case is an example of a mode of onset, namely, stumbling gait, very apt to be overlooked.)

8. *Myra M.*, aged 8 years 7 months. (April 22nd to May 26th.) (Dr. Cheadle's case.)

Has been working hard at school for six or eight months ('overworked,' says the mother). Four months ago she woke up screaming, and for a time did not recognise her mother. But nothing further was noticed until six weeks ago, when limb movements and defects of hand grasp were observed. *She was kept at school, notwithstanding, for three weeks longer,* during the latter part of which time her speech began to fail, and she became 'stupid.' A moderate general chorea; making good recovery.

The points to be specially noticed in the foregoing list are these:—

1. Only one of the number is a boy, and with him a special alarm was a contributing cause.

2. There was school worry in all the cases except one (No. 5), where there was the injurious excitement of performance at a school concert.

3. In no less than five the children were kept at school after their limb disorder had been noticed.

The way of St. Vitus' Dance, when school-made, is commonly this: At first the temper deteriorates, the child gets petulant, fretful and capricious, has restless nights and often headache, and if these signs are not read aright and school work is not relaxed, there arises presently bodily restlessness. Even this may be overlooked, because children are by nature restless. But teachers who are on the watch and already warned by the symptoms just mentioned, will easily see that this overmovement is of a new kind and not merely restlessness. It will often apply, particularly at the first, to one side or one limb, with a general want of control; this is expressed most in one place, very commonly in one hand.

The sad consequence to the child of an affection such as this so long as it is unrecognised is obvious. Infirmity provokes punishment and punishment aggravates infirmity. Bad writing and untidy sums are met by scolding and slaps on the back; these correctives in their turn produce worse writing and worse sums, until at length by a self-acting process the disorder grows into notice and the child is put to bed. School-made chorea has in fact two stages: the teacher's and the doctor's. At first, while at school, it is subjected to penalty, and at last, when by such correction it is fully developed, it is remitted to the doctor and allowed time for recovery.

A little discernment and some quite elementary knowledge

on the part of teachers would not only save much undeserved suffering, but also enable them in many instances to arrest at an early stage the progress of a disorder they are now largely chargeable with promoting. To this end the following points for observation are suggested.

1. Movement disorder in children, the index and product of some form of school worry, may be known by nice observation of the higher muscles (the face and hands) before it has reached the stage to which the term chorea or St. Vitus' dance is usually applied.

2. Among the incidents of school life apt to be injurious there stand out prominently: competitive examination; moving into a higher class; sums (too difficult or ill-explained); punishment; the ridicule and ill-treatment of older children.

3. When school children (and especially girls) alter in temper, do lessons less well and less willingly than usual, get untidy, careless and slovenly, write (and perhaps read) ill, or drop their books and slates; in a word degenerate in mind and body, enquire of the mother as to the home conduct and temper. Ask especially how the child sleeps, whether she complains of headache (or limb-ache) and whether her food is sufficient.

4. The best index of muscular infirmity tending to St. Vitus' Dance is *the hand*. Face mobility may be mere nervousness; the tongue may be tremulous by nature; the hand test is infallible. It is thus applied: bid the child hold up both hands open with extended arms and the palms towards you. If that be done steadily, both hands upright and both alike, no finger or thumb quivering, no falling back or hanging forward of either hand, nothing to choose between the position of the two, then the child may be acquitted of chorea. This test may be confirmed by another. Let the child place its open hands upon yours, palm to palm. Looking then at the backs of the

child's hands, during two or three minutes, note whether fingers and thumbs (and especially the latter) repose naturally, without tremor and without restraint.

5. When a child returns to school on recovery from chorea, have in mind the liability to relapse, make allowance in school work, and be specially watchful of faulty muscular conduct.

## CHAPTER VI.

## PATHOLOGY.

Early views—Connection with rheumatism—with spinal meningitis—with pericarditis—with enfeebled nutrition—Embotic theory—Dependence on vascular and nervous changes—On disturbed nutrition of cerebral ganglia—On altered states of blood—On functional disturbance—Continental opinion—Retrospect.

I PROPOSE to set down in this place, in something like chronological order, a short summary of the several hypotheses as to the pathology of chorea which have been put forward since the time that the disease was first accurately described.

The connection of rheumatism with chorea was first asserted at the beginning of this century. Dr. Bright mentions that in the 'Syllabus of the Practice of Medicine,' published at Guy's Hospital in 1802, 'rheumatism is distinctly mentioned as one of the exciting causes of chorea.' He adds that in later editions of the same work (as in that of 1820), chorea is said to alternate with acute rheumatism, 'but through what organ or by what intervention it occurs is not conjectured.' In the following year, and independently, as he expressly asserts, of these earlier observations, Dr. Copland drew attention to the rheumatic origin of chorea, upon the evidence of a case which he quoted of 'rheumatic pericarditis and disease of the membranes of the spine, with which chorea alternated.' This change from rheumatism to chorea he sought to explain by 'metastasis from the joints to the membranes of the spinal cord.' 'In nearly all the cases,' he writes, 'there has been a

marked disposition of the rheumatic inflammation to recede from the joints and attack the internal fibrous serous membranes as those of the cerebro-spinal axis and the pericardium.<sup>1</sup>

A more precise reference to the pericardium in association with chorea occurs in the paper by Dr. Bright, from which I have just quoted, on 'Cases of Spasmodic Disease accompanying Affections of the Pericardium' (*Med.-Chir. Trans.*, vol. xxii.). In this communication the author dissents from the conclusions of Dr. Copland connecting chorea with spinal meningitis, and expresses his belief that a much more frequent cause of the complaint is pericarditis, 'although in some instances the coverings of the cerebro-spinal mass might be implicated.' From the inflamed pericardium, as Dr. Bright supposed, 'an irritation was communicated to the spinal cord,' of which irritation convulsive movement was the consequence.

It is to be remarked, however, that Dr. Bright's cases, as described by himself, are illustrations of spasmodic disease rather than of chorea. In one of them there were epileptic seizures and tetanic spasms; in another, the symptoms put on the 'character of most violent convulsions,' 'more violent,' it is added, 'than is almost ever seen in chorea.' Both these cases were fatal. In some of the other examples, too, 'there might be doubt,' as Dr. Bright admits, 'of the correctness of the diagnosis.' What his paper seeks to make out is indicated by its title—the connection, namely, between violent and often fatal muscular spasm and pericarditis, the patients not being children but young men and women.

In order to show how little such views found acceptance in this country, and how vague and indefinite was the pathology of chorea more than ten years later, Dr. Todd's opinion may be next quoted. 'It is easier,' says this physician (in lectures published in 1854, but delivered some time before), 'to say

<sup>1</sup> See Copland's Dictionary, art. 'Chorea.'

what chorea is not, than to describe what its essential nature is. We may regard it as a disease which depends upon a debilitated state of the system, which does not in any way arise from an inflammatory or hyperæmic state of any part of the great nervous centres or of other organs. Almost without exception, in those cases which terminate fatally, we fail to detect any morbid alteration which physiologically could give rise to the phenomena. If I were to refer to any particular part, it would be to that which may be regarded as the centre of emotion. The remarkable frequency with which the attack has been traceable to fright as its cause points clearly to this part of the brain as the *primum movens* in the production of choreic convulsions. The chain of phenomena would then be as follows: first, a peculiar diathesis; secondly, a more or less enfeebled nutrition; thirdly, a strong mental impression which disturbs the centre of emotion, and through it deranges the action, more or less, of the nervous system and of a corresponding portion of the muscular system.’<sup>1</sup>

It was eleven years after Dr. Bright’s paper, namely in 1852, when the reality of the connection between rheumatism and chorea had been recognised by text books as well as the occurrence, in the latter affection, of systolic murmur, that Dr. Kirkes wrote his celebrated essay on embolism, destined to exercise an important influence upon pathology. It was entitled ‘On some of the principal effects resulting from the detachment of fibrinous deposits from the interior of the heart and their mixture with the circulating blood.’<sup>2</sup> Towards the end of this paper occurs the passage which may fairly be regarded as the origin of what is now called the embolic theory of chorea. ‘I would suggest,’ writes Dr. Kirkes, ‘that many functional disorders of the nervous system, especially

<sup>1</sup> Todd’s *Clinical Lectures*, ‘Nervous System,’ p. 442.

<sup>2</sup> *Med.-Chir. Trans.*, vol. xxxv.



chorea, may be thus explained [*i.e.* by cerebral embolus]. The frequent existence of a cardiac murmur in chorea, and the presence of warty vegetations on the valves of the heart so commonly found in fatal cases of the disease, are in favour of such a view.'

Of the development of this doctrine, and the mode in which it is now expressed, Dr. Hughlings Jackson may be taken as the exponent. 'I suppose,' says this pathologist, 'that the excessive movements occurring either in chorea, or in epilepsy, or epileptiform seizures, are produced by discharges of grey matter, which, except for great instability from over-nutrition (not better nutrition), is healthy. We cannot expect to discover, with our present means of research, the alterations in grey matter on which excessive discharges depend.' 'My opinion is,' he goes on, 'that the direct pathological state leading to instability of grey matter, producing choreal movements, is increased quantity of blood in the periphery of the capillary district embolised.' 'Of course,' he adds, 'this opinion is hypothetical, and so is every one's opinion as to the nature of lesions insusceptible of anatomical demonstration.'<sup>1</sup> As regards the seat of these lesions, Dr. Hughlings Jackson, following Russell Reynolds, fixes upon the corpus striatum, insisting that there is frequently disease of that part of the brain which superintends the movements of the tongue in uttering syllables. He asserts, moreover, 'the frequent one-sided nature of the movements of the limbs, and *their often dying out into definite hemiplegia*' (words which I venture to italicise), 'as pointing to disease at or near the corpus striatum.'<sup>2</sup>

It has not escaped Dr. Hughlings Jackson that the special character of choreic movement as contrasted with the convulsion of epilepsy needs some apology. 'The elaborateness of

<sup>1</sup> *British Medical Journal*, Dec. 23, 1876.

<sup>2</sup> *London Hospital Reports*, 1864.

the movements,' he says, 'is strong warrant for the inference that the changes causing them must be seated in the brain and not in the cord'; and he thinks it 'most probable that the convolutions are the parts diseased.'

From views thus mainly hypothetical we may turn to those of Dr. Dickinson, arrived at directly from *post-mortem* evidence, and which, in the opinion of their opponents, only fail when the attempt is made to apply them to the actual symptoms of the disease in question.<sup>1</sup> It is impossible in this summary to reproduce an adequate account of the facts upon which Dr. Dickinson depends for his conclusions. These facts are based mainly upon a minute examination of seven fatal cases of chorea, or, as I should prefer to describe them, fatal cases *with* chorea, in three of them at least the choreic movements being of no great severity.<sup>2</sup> The morbid changes, described as remarkably constant in kind and place, are thus summed up by the author. 'In *place* the changes affected both brain and cord. As regards the two sides of the body they were either exactly symmetrical or tended to be so. The parts of the brain most amenable lay between the base and the floor of the lateral ventricles in the track of the middle cerebral arteries; the substantia perforata, corpora striata, and the beginning of

<sup>1</sup> *Med.-Chir. Trans.*, vol. lix., p. 15, 'On the Pathology of Chorea.'

<sup>2</sup> Thus in Case 1, a girl of 10, the child is described as restless and sleepless, her movements much increased on the 21st, on which evening, 'after acute pains in the lumbar region, she somewhat unexpectedly died;' there were vegetations on the aortic as well as the mitral valves. In Case 5, a boy of 11, the use of the limbs and the power of speech were slowly restored, and the boy was sent into the country convalescent. There 'his heart symptoms became more pronounced, without any return of those of chorea.' He died of his heart disease. In Case 6, a girl of 13, 'the symptoms were not apparently threatening, and the chorea in particular obtained little attention.' She, too, died suddenly, and had both mitral and aortic disease.—*Vide loc. cit.*, pp. 1 *et seq.* A similar remark will apply to some of the fatal cases of chorea by other authors. See *e.g.* H. Jones, *loc. cit.* p. 560.

the Sylvian fissures. In *kind* the changes were all connected with vascular disturbance. The injection was general to all the vessels, most marked in the arteries; when the sources of hæmorrhage could be determined they were always arterial; the degenerations were usually periarterial, and the spots of sclerosis similarly placed. The first visible change would seem to be the injection or distension of the arteries, succeeded by extrusion of their contents to the irritation and injury of the surrounding tissue.'

'Of the cord no region was exempt; but perhaps the cervical and dorsal regions were usually more affected than the lumbar.' Both white and grey matter 'shared in the vascular distension. This condition, however, was usually most marked in the vessels belonging to, or in connection with the lateral part of the grey matter about the root of each posterior horn. This was also the chosen situation of the more definite and special changes.' In commenting upon these changes as a whole, the writer observes: 'The nature and steps of the morbid process are open to view, hyperæmia, exudation, and its consequences, but not so the causes in which the series has taken origin. Arterial repletion seems mainly concerned in the development of the disease.'

As regards embolism, the author remarks not only upon the absence of any signs of such impaction, but upon the constancy with which the changes repeated themselves in certain portions, and the equality with which they affected both sides of the body as conclusive objections to that hypothesis.

'The results,' says Dr. Dickinson in conclusion, 'in muscular excitement rather than in paralysis or loss of sensation, may be associated with the character of the lesions, which are points of irritation rather than planes of section, and as such calculated to produce irritation rather than paralytic effects, not so much to cut off, as unnaturally to excite nervous

function.' The author alludes to the similarity between the morbid changes of chorea and those of diabetes, and he concludes with a bold attempt to bend clinical facts into some compliance with the demands of morbid anatomy. 'Every period of life has its own regions of nervous susceptibility; in childhood the motor; in adolescence the emotional; in advancing years the mental, and coevally, or nearly so, that part of the nervous mechanism which instigates glycosuria. Much the same mental impression may make a child choreic, a girl hysterical, or a man diabetic. And thus both in external origin and in the nature, though probably not in the site of the organic changes, we see resemblances and alliances between nervous disorders which in their symptoms betray little similarity.'<sup>1</sup>

In noticing these observations of Dr. Dickinson the advocates of embolism maintain that the condition just described is not incompatible with that theory. 'The local periarterial softenings,' Dr. H. Jackson suggests, 'account for the choreal paresis,' while the cerebral changes described are not very unlike those producible by emboli, regard being had to size of arteries plugged.' He admits at the same time that the embolic hypothesis supposes the cord to be unconcerned in the irregular movements of the disease, and even confesses that, as a matter of fact, the absence of embolism is much more common than its presence.

Dr. Bastian,<sup>2</sup> supporting in some respects the conclusion of Dr. Dickinson, and relying with him upon anatomical evidence, agrees with Dr. Hughlings Jackson in attributing chorea to disturbed nutrition in the corpora striata and adjacent parts. In place of embolism, however, he would put thrombosis. He points out that the production of multiple

<sup>1</sup> *Loc. cit.* p. 39.

<sup>2</sup> *British Medical Journal*, Jan. 20, 1877.

minute thromboses 'might easily give rise to minute vascular injections,' but is careful to add that 'other hyperæmias, more or less secondary in their duration and mode of origin, may easily be confounded *post-mortem* with those which are hypothetically supposed to lie at the root of chorea.' In support of this opinion he refers to three cases of his own, where, with hyperæmia of the ganglia at the base of the brain, and especially the corpora striata, occlusions were found in the small vessels of these parts.<sup>1</sup> He sums up his view of the etiology and pathology of chorea: 'I look (certain rare cases excepted) to an altered and often anæmic blood state as its predisposing cause in individuals of a certain age and nervous temperament. Secondly, I look to the initiation in such individuals of a disturbed nutrition in the corpora striata and adjacent parts of the brain, tending to issue and often actually issuing in what may be called a subacute inflammation of these centres, often characterised in part by the production of multiple minute thromboses.'<sup>2</sup>

In harmony with these views, so far as regards locating chorea in the corpus striatum and optic thalamus, and in partial agreement with them as accepting capillary embolism of these parts and of their vicinity as one of its causes, we may notice next the opinions of Dr. Broadbent. In addition to embolism, however, Dr. Broadbent<sup>3</sup> would recognise many other sorts of disturbance of nutrition of the cerebral ganglia, 'as, for instance, peripheral influences arresting the reflex process, direct lesions of the ganglia due to fright or to mechanical lesions,' as possible causes of chorea. 'The processes of chorea are in fact such as weaken the force of the nervous apparatus without destroying its structure. Hence the weakness of the

<sup>1</sup> *Ibid.*, July 13, 1877, p. 38.

<sup>2</sup> *British Medical Journal*, Jan. 20, 1877.

<sup>3</sup> *Ibid.*, April 17-24, 1869.

muscular force and diminished sensibility so common in chorea. Hence also its frequent termination in paralysis.<sup>1</sup> 'The functions of a particular part of the brain are deranged ; chorea is a symptom, not a disease.' To the condition of system of which it is the consequence, Dr. Broadbent would give the name of 'delirium of the sensory motor ganglia of the brain.'

Having thus reached in our search after the physical basis of chorea, the point at which it is affirmed that the nervous apparatus is *weakened*, but not destroyed, that the disorder is a symptom, but not a disease, and that the condition of the nervous ganglia concerned is best described by such a word as *delirium*, we may fairly conclude that anatomical hypothesis has pretty well exhausted itself.

Such a conclusion is as good as affirmed in the words of Dr. Ross,<sup>2</sup> in his elaborate work on 'Nervous Diseases.' 'The lesion, whatever it be, is a widely spread one, and is not limited to the basal ganglia or any other special part of the nervous system.' Dr. Bristowe speaks to like effect.<sup>3</sup> 'It is not known,' he admits, 'what parts of the central nervous organs are the seat of the disease, or what is the nature of the morbid process going on in the affected parts.' Observation of the disorder of respiration and of heart action incline him 'to add the medulla to the other parts implicated, while the resemblance of the affection to locomotor ataxy suggests involvement of the cord as well.' Furthermore, in allusion to Dr. Dickinson's views, Dr. Bristowe says in conclusion : 'The tendency which the vascular changes have

<sup>1</sup> The frequent termination of chorea in paralysis, asserted both by Broadbent and Hughlings Jackson, is, I submit, a misleading expression, and ill-describes the partial short-lived paresis of the affection.

<sup>2</sup> Vol. ii., 783.

<sup>3</sup> 'Practice of Med.,' 1120.

to induce sclerosis will explain the wasting of muscles, rigidity of limbs, and permanent paralysis which occasionally complicate chorea or supervene upon it.'

These expressions, it must be insisted, not only fail to supply any definite pathology for chorea, but in their efforts to do so, they altogether misrepresent its clinical characters. Wasting of muscles, rigidity of limbs, and permanent paralysis are all highly exceptional in chorea, while its claims to be regarded as a functional disease, whether right or wrong, are put in their strongest light by considering its utter unlikeness to locomotor ataxy.

But when physical changes are lost sight of there still remains a wide field for conjecture outside morbid anatomy. We leave the tissues and make appeal to the blood. We have seen already (p. 66) how the condition of anæmia has been invoked to account for the heart symptoms of chorea. But Dr. John Ogle,<sup>1</sup> with whom Dr. Hammond,<sup>2</sup> of New York, fully agrees, would give a wider rôle to the influence of altered blood 'He is led to regard fibrinous deposits on the heart valves as results of some antecedent condition of the blood, common also to the choreic condition.'

From conjectures like these the transition is easy to a pathology which abandons all attempts at anatomical description and speaks in general terms of 'altered cerebral nutrition' and 'abnormal conditions of nerve discharge,' whose seat 'may with probability be ascribed to the motor region of the cerebral cortex.'<sup>3</sup> Thus Dr. Robert Barnes,<sup>4</sup> considering chorea in its connection with pregnancy, writes: 'The con-

<sup>1</sup> *Med.-Chir. Review*, January, 1868.

<sup>2</sup> 'Diseases of Nervous System,' p. 614.

<sup>3</sup> See Fagge and Pye-Smith, vol. i., p. 749.

<sup>4</sup> Dr. Charlwood Turner, in a striking communication to the Pathological Society, May 17th, 1892, comments on some sections he exhibited from the Rolandic region of the brain in five cases of death from chorea,

dition upon which the latent disposition to chorea depends is some change of nutrition or of structure of the nervous centres unimportant under the ordinary conditions of life, but liable to be called out into renewed activity under that special increase of central nervous development which is the constant attendant upon pregnancy.<sup>1</sup> The chorea of puerperal women Dr. Barnes believes to be almost always a revival of the same disease in childhood, having pregnancy for its immediate exciting cause, in virtue of 'the exalted central nervous irritability' of that condition, which is thus 'a test of the soundness of the nervous system.'

As nearly in accord with these views, reference may be made next to the opinions of Dr. Handfield Jones; and here, even more than in the previous instances, comes the difficulty of doing justice to the author in such curtailed statement as the present design necessitates. The following quotation, however, gives in Dr. Jones's own words a summary of his opinion. For its illustrations I must refer to his well-known work.<sup>2</sup>

'The view which I take of the pathological events in chorea is the following. The motor centres especially, and also not unfrequently the intellectual, emotional, and sensory, in persons of weak organisation, fall into a state of paresis, either in consequence of a shock or more gradual injury, or of some toxic matter in the blood, or of peripheral irritation, all of which may generate the same peculiar condition. The paresis in all these parts may take the form of hyper-excitability or of paralysis, the former being much the more frequent. The nerve exhaustion, aggravated more or less by the jactitation, showing lesions of some of the large pyramidal cells of the cortex. He infers thence that the phenomena of the disease are due to such lesions producible by disturbance of the nutrition of the brain from various causes.'—*Trans. Pathological Society*, 1892.

<sup>1</sup> *British Medical Journal*, Dec. 9, 1876.

<sup>2</sup> 'Functional Nervous Disorders,' p. 361, *et seq.*



involves the vaso-motor nerves of the cerebral and spinal arteries especially, and conditionates relaxation of their muscular walls as well as impairment of the tone of the capillaries and hæmorrhage. These, however, are of course not necessary events, even in fatal cases. Occasionally actual inflammation of the nerve-centres results from the hyperæmia. Pulmonary congestion and consolidation may be produced in the same way, and possibly, in some instances, valvular lesions of the heart. This view, that chorea is essentially a functional disorder of the motor centres, is in harmony with its frequency in children and females, whose motor apparatus is more prone to be weak and irritable, with its affinities to epilepsy, hysteria, paralysis, and insanity.'

As for the connection between chorea and rheumatism, it seems to this observer to be best explained by regarding the motor disorder in just the same light as we do delirium, to which it has much affinity. 'In typhoid we have an intestinal lesion, and commonly delirium; but we do not assume a connection between these, but consider the specific poison to give rise to both. In rheumatic fever we have also often a lesion (cardiac) and delirium, both as co-products of the cause of rheumatism, not one as the cause of the other. If we substitute motor disorder for intellectual, why need we change our view?'

Before concluding this statement it still remains to notice some of the theories and researches of continental pathologists. Yet any detailed account of these may be excused after the admission of Ziemssen, that 'the sum of knowledge of the pathologico-anatomical changes of chorea is so small that it is scarcely of consequence to attempt to explain it.'<sup>1</sup> It is admitted that England and France have been the chief contributors both to the statistics and pathology of the affection,

<sup>1</sup> Ziemssen, 'Cycl.' *loc. cit.* 457 *et seq.*

while in Germany the observations of English pathologists upon the embolic origin of chorea and its connection with endocarditis have attracted more attention than any investigations of their own. It is due, however, to the high authority of certain continental pathologists that their contributions to this subject should be referred to shortly.

Rokitansky, in 1857, asserted the occurrence in chorea of interstitial connective-tissue growth in the central nervous system; and Steiner, ten years later, confirmed the observation in a single case, and so far as it concerned the spinal cord. He found also, in two cases, serous effusion within the spinal canal, and in one, hæmorrhage at the upper part of the cord at the exit of the nerves. Similar changes, so far as the cord is concerned, together with hyperæmia of the medulla and spots of softenings, have been now repeatedly met with. Yet it is to the brain, rather than the cord, that continental observers, influenced like our own by considerations of physiological necessity, have chiefly looked to discover the structural basis of chorea.<sup>1</sup> Cerebral changes, it is asserted, affect the brain cortex and grey substance of the great basic ganglia; but their precise character, far from being exactly stated, is made to rest rather upon clinical than anatomical observation. As with the cord, the brain changes are said to include 'interstitial hyperplasia, with regressive metamorphosis of nerve elements, sometimes confined to one hemisphere or its great ganglia, and sometimes diffused over the whole brain, preferring the grey substance, and extending always to the cord, and even the peripheral nervous system.'<sup>2</sup>

Again, Charcot has recorded some observations upon cases of hemichorea after hemiplegia, in which cicatrices were found

<sup>1</sup> Ziemssen, *loc. cit.*, p. 460.

<sup>2</sup> As regards the latter, the observations of Elischer may be quoted. He found the changes just described for the cord equally marked in the peripheral nerves, the connective tissue between nerve bundles being greatly

at the posterior end of the optic thalamus, and (in two cases) of the anterior corpus quadrigeminum of the affected side. In other instances, where hemiplegia followed chorea, the same observer found a blood clot as large as a nut in the posterior half of the optic thalamus. In accepting the evidence of such cases, however, referring chiefly to adults, we are transgressing the proper limits of the subject, and complicating the pathology of chorea with that of paralysis.

A similar objection applies to many of the other quoted cases, where it will be found that the chorea is combined with some other affection, as with mania or encephalitis. It is admitted, indeed, by Ziemssen and others—and I shall presently quote instances of this kind—that ‘chorea may occur as a transitory phenomenon in the course of cerebral lesion, and presently give way to other and severer disturbances.’<sup>1</sup> (Case 16.) It is misleading to adduce the *post-mortem* appearances in such cases by way of illustrating the anatomical changes proper to chorea.

Lastly, experiments have been made upon animals by Chauveau and others,<sup>2</sup> and it has been shown that the muscular twitchings of dogs persist after section of the cervical cord, and, further, that these may be produced or increased by mechanical irritation of the posterior columns. But limb twitching is not chorea nor even a symptom of it. Thence it is concluded that the limb-twitching depends on the cord, and not on the brain.

developed, and the medullary sheaths of the nerve fibres tumefied, while their axis-cylinders had disappeared. The nerves examined (the median and sciatic) seemed to the naked eye abnormally small and flat.

<sup>1</sup> *Arch.-Gen. de Méd.*, March, 1866, 455.

<sup>2</sup> In this country Dr. Angel Money made some experiments upon animals with a view to producing the symptoms of chorea by injecting foreign material into the cerebral vessels, but on his own confession the attempt was a failure. *Brit. Med. Journ.*, July 17, 1886.

In reviewing the several observations and conclusions which have now been enumerated, it is easy to see that the theories in reference to chorea which have successively gained acceptance represent so many attempts to accommodate this particular affection with the pathological doctrines of their day. When first emerging from the obscurity of humoral pathology, chorea, partly no doubt in virtue of the pain that often attends it, is found in some sort of alliance with rheumatism. Presently, upon the observation of its occasional association with acute rheumatic arthritis, the prevalent belief in metastasis is appealed to, and the muscular disorder is accounted for by an inflammation transferred from the joints to the spinal cord. Next, and following the researches of Marshall Hall upon the functions of the cord, we find the principle of reflex irritation made use of to explain the dependence of chorea upon pericarditis and intestinal irritation. At a still later period, and when further observations of the heart symptoms of chorea had made the application easy, comes the discovery of embolism, both to explain the cardiac disturbance and to satisfy those phenomena of the disease which better study had shown to depend upon the brain rather than the spinal cord.

But if the pathological history of chorea has been one of revolution, its clinical history (in so far as the two may be separated) has been one of continuous progress; and it has come to pass with the gradual accumulation of facts, that while precise and definite hypotheses have been found faulty and insufficient, the only pathology that survives hardly goes further than to translate its symptoms into physiological language. We have mere definitions of the disorder which are unassailable, which venture nothing and explain nothing; but what is wanted is some explicit statement of the anatomical conditions on which chorea depends which may be brought into harmony with its actual phenomena.

## CHAPTER VII.

## THEORIES IN THEIR APPLICATION.

Clinical conditions to be satisfied—The symptoms of embolism compared with those of chorea—The direct evidence in favour of the embolic theory—Perivascular degeneration of the brain and cord as a cause of chorea—Hypothetical explanations—Chorea ascribed to temporary trophic changes—Application of such theories—The ataxy of chorea.

I TURN then once more to chorea itself, and would place side by side with it the several hypotheses which have just been enumerated. No theory can be finally accepted which fails to satisfy all the conditions of the problem. The main features of chorea are obvious and beyond dispute, as that it is a child's disease, that it prefers the female sex, that it is apt to recur, that it affects the hands especially, and the arms and face more often than the legs, that it is more often both-sided than one-sided, that complete recovery is the common rule, and death or any permanent disablement the rare exception, and so forth. Whether, therefore, it be alleged that embolism, or thrombosis, or perivascular softening, or 'disturbance' in some motor centres, or 'toxic influence' is at the root of the affection, each of these assumed conditions needs for its justification to be brought into relation and harmony with the known symptoms of the disorder it professes to explain.

Now, if this comparison be fairly made I think it will appear :—

1st. That there is no morbid condition as yet anatomically described which, so far as we have independent knowledge of

it, is capable of producing symptoms at all similar to those of chorea.

2nd. That the hypotheses which, apart from demonstration, really provide the best for the actual phenomena of chorea, do not in fact present any distinct picture of a definite morbid change.

3rd. That having regard to the special characters of chorea, it is difficult to conceive of any morbid condition whatever, anatomically demonstrable, calculated to produce it, or indeed, compatible with it.

These propositions may be taken in their order:—

I. *No morbid condition as yet known produces symptoms at all like chorea.* The morbid anatomy of the disorder, or as much of it as has survived, begins, as we have seen, with its association with endocarditis,<sup>1</sup> an association soon to be supplemented and explained by the observations of Dr. Kirkes upon embolism in 1852. Taking, therefore, this, the most concise and ingenious of all the theories having a purely anatomical basis, we may enquire, in the first place, whether embolism, so far as we have independent knowledge of it, is capable of producing symptoms at all similar to those of chorea.

Now, the ordinary symptoms and course and cause of embolic obstruction as regards the brain are known. Making

<sup>1</sup> In speaking of the several anatomical conditions which have been definitely described as representing the material expression of chorea, it is unnecessary to refer again to doctrines now abandoned. Yet the observations of Dr. Bright in 1841 upon the connection between pericarditis and chorea have this singular value; they point to a connection which, although different in form from that which Dr. Bright asserts, is really implied in the evidence which his paper affords. There is reason to believe, as we shall presently see, that severe and fatal choreic attacks are sometimes in direct relationship with endocarditis. It is in the highest degree probable that Dr. Bright's cases, although referring expressly to pericarditis, are in fact illustrations of this connection, inasmuch as in acute rheumatism the endocardium and the pericardium commonly suffer inflammation together.

allowance for great diversity in the response to cerebral injury, as well as for modifications due to the extent, size, and site of the plugging, our actual information under these headings may be summed up generally as follows:—(a) For *symptoms*: a sudden vertigo or an acute head pain, with faintness or sickness; a more or less complete hemiplegia; impaired intelligence and sensation; and, if the hemiplegia be right-sided (*as it most often is*), either aphasia or inability to utter articulate sounds. (b) For *course*: a gradual but interrupted decline, or an incomplete and precarious recovery; nutrition changes in the muscles of the affected side; a liability to recurrence of the first fit; often a fatal termination by way of coma. Such or such like clinical features of embolic cerebral obstruction are accounted for by reference to its morbid anatomy. Its usual seat, mainly determined by the distribution of the middle cerebral artery, is the left rather than the right side of the brain, while secondary softening is an early consequence of the original accident. (c) For *cause*: the conditions which belong to degeneration; to endocarditis; to acute rheumatism; and to the puerperal state.

Such is the general description of cerebral plugging, so far as it has been observed clinically and anatomically; and if the account have reference to minute embolism, we have but to add that there is often active delirium, high temperature, and a train of symptoms, rapidly fatal, resembling specific fever.<sup>1</sup> How far does all this apply to chorea; to a disease confined almost to childhood; favouring the female sex; having no preference for the right side; seldom sudden in access; hardly ever exhibiting true aphasia, high temperature, vertigo or sickness; producing no special wasting of the affected muscles; implying hardly any danger to life or limb, and apt

<sup>1</sup> See, for example, two cases of cerebral embolism reported by Dr. Dickinson, *British Medical Journal*, May 21, 1881, p. 795.

to disappear at that precise period of life when, with the greater frequency of heart disease and the setting in of degenerative changes, the liability to embolism and thrombosis is largely increased? Putting together all that we know of the plugging of cerebral vessels, large or small, in its symptoms, associations and subjects, it is directly opposed to all that we know in respect of chorea.<sup>1</sup> If it could be shown, as it can not, that this particular morbid condition was constantly present in cases of chorea, the observation would offer a new difficulty; we should still have to look elsewhere for the explanation of its symptoms.

But we have yet to inquire upon what ground of actual observation this doctrine of vascular plugging depends. 'I am aware,' says a very candid and fair advocate<sup>2</sup> of this belief, 'that the morbid anatomy of chorea is not considered to lend much support to the attractive hypothesis of embolism.' Drs. Wilks and Moxon have never met with microscopic emboli in any instance. In one case where, together with chorea, hemiplegia had followed an embolus, 'there were no discoverable emboli in the small vessels.' These authors are unable, indeed, to recognise *any* constant morbid appearance after death in chorea. Similarly Dr. Gowers and Dr. Ferrier have failed to discover emboli after diligent search. It is not too much to say that instances of minute embolism in this association are conspicuous by their rarity. Cases of the kind, as those of Dr. Broadbent and of Dr. Tuckwell,<sup>3</sup> are

<sup>1</sup> The connection of chorea with acute rheumatism and with child-bearing may be alleged to the contrary. But the rare occurrence of embolism in both these conditions concerns the large vessels, and not the capillaries, while the chorea of child-bearing, as Dr. Barnes has pointed out, is almost always but the revival of a former chorea in childhood.

<sup>2</sup> Dr. S. Mackenzie, *British Medical Journal*, Dec. 23, 1876.

<sup>3</sup> *Medical Times and Gazette*, 1875, p. 482; *British and Foreign Medico-Chirurgical Review*, 1867; *St. Bartholomew's Hospital Reports*, vol. v.



quoted over and over again, but it is admitted that they conflict with the general experience. 'I have never seen an instance,' says Dr. Dickinson, 'in which the well-known blocking [of embolism] as found after death, has been conjoined in life with choreic symptoms' (*loc. cit.* p. 36). If this theory of vascular plugging is to be established, it must be, as Dr. S. Mackenzie admits, 'upon evidence of a clinical and physiological nature.' Where is such evidence to be found? It would appear, on the contrary, that the condition of minute vascular plugging, whether of embolism or thrombosis, is not only insufficient theoretically to account for the special phenomena of chorea, but that, as a matter of fact, it is only exceptionally met with in connection with it.

Let it be granted, however, that 'an increased access of blood to a district which is embolised but otherwise healthy,' is, by whatever agency, the cause of chorea, is such a condition really compatible with the phenomena it professes to satisfy? I maintain that it fails, both in respect of embolism and in respect of increased access of blood. Our knowledge of cerebral embolism assures us that grey matter improperly nourished soon disintegrates and softens. Allowing, therefore, that this result of embolism would give rise to chorea first, it would give rise to paresis afterwards. Choreia would not be the recovering disease which we see; it would be intimately allied with paralysis. Else the hypothesis which provides embolism as its cause, must provide something besides, which shall presently dispose of such embolism.

But further, and apart from this difficulty, upon what observation is the hypothesis based that hyperæmia of grey matter occasions chorea? In the many forms of intracranial growth, syphilitic, tubercular, and sarcomatous; in aneurism; in meningitis; in the changes which precede softening; in the reparative processes after cortical hæmorrhage, a localised

hyperæmia of grey matter may be assumed to arise at some time or other. The symptoms of these several conditions, as is well known, present the greatest variety. They include almost every kind of nervous disturbance except chorea; spasm, whether tonic or clonic, 'limited to the distribution of a single motor nerve or implicating a group of muscles on one side of the body;' emotional disorders resembling hysteria so closely as to be mistaken for it; incoherence, mania, delirium, obstinate vomiting, acute pain, aphasia, but never (or hardly ever)<sup>1</sup> chorea. Especially is it to be remarked in the case of meningitis, a disease which both involves hyperæmia of grey matter and has children for its favourite subjects, that with all the variety in its symptoms chorea has no place. There is nothing more remarkable than the way in which this movement disorder keeps itself separate, not only from all those brain diseases which imply active trophic change but from all active disease whatever.

Similar remarks would apply to the nervous changes pointed out by Dr. Dickinson, and shortly described in the last chapter. Although this condition, consisting in spots of perivascular degeneration distributed symmetrically throughout the brain and cord, was found, not only in chorea but in diabetes as well, it is obviously not the kind of change we are seeking, and, being found, it is impossible, without a revolution in physiology, to reconcile with it the actual phenomena of chorea. How can these periarterial spots, symmetrically arranged, affecting the cord as well as the brain, and tending to ultimate destruction of nerve substance, be compatible with a disorder which is transitory and sometimes one-sided, and which almost always leaves the functions

<sup>1</sup> I make this reservation in view of those individuals who have already suffered chorea in childhood, and with whom, as is well known, many provocations, and cerebral disease amongst the rest, will serve sometimes to revive the same affection.—See Case 16, Chap. XI.

of the cord unimpaired? How, indeed, can those who appeal exclusively to anatomy to explain disease be content to accept one set of changes for affections so diverse as disseminated sclerosis, diabetes, and chorea?

The difficulty has been felt and expressed. And accordingly, in place of a pathology which claims to have the direct sanction of anatomical observation, we are offered hypotheses expressly framed to meet the conditions of the case, but which, while still insisting in part upon a physical change, eke out the rest by means of some of those general expressions imputing blame to the blood or the vessels which are so ready of service where demonstration fails. I have already alluded (p. 90) to Dr. Bastian's opinion that chorea is due to 'disturbed nutrition' of the corpora striata, 'issuing often in sub-acute inflammation of those centres, and characterised in part by the production of minute multiple emboli, a condition originating in an altered state of blood.'<sup>1</sup> So far as the anatomical part of this description is concerned, Dr. Bastian himself calls attention to the scantiness of its direct evidence, and admits the possibility of confounding what 'lies at the root of the disease with other hyperæmias more or less secondary in duration and mode of origin.' And for whatever is beyond anatomical description, there are difficulties in the way of accepting what is asserted, arising partly from its want of definiteness and partly from its want of truth. 'Disturbed nutrition' is hardly a tangible expression, while any theory which needs the supposition of a blood-change is at once negatived by clinical facts. Chorea attaches to no special blood-state; it occurs to the plethoric as well as the anæmic; and all the 'blood-poisons' that excite pyrexia serve rather to suppress than to cause it. Even in the case of acute rheumatism, which would seem at first sight to give shelter to some such theory, it is not until the

<sup>1</sup> *British Medical Journal*, p. 67, 1873.

active febrile stage of that disease has disappeared that the chorea comes.

And so by degrees, with the gradual substitution of hypothesis for observation, we come at last to theories which are *all* hypothesis, a mere writing out of the symptoms of chorea in technical phrase, where, although some morbid change or other is still implied, no attempt is made to reconcile the actual phenomena with any definite localised lesion. It is felt, indeed, that this task of reconciliation becomes the more perplexing just in proportion as anatomical description becomes the more explicit. It is easy to accommodate chorea to general expressions of altered nutrition and disturbed motor centres; it is most difficult to accommodate it, even in theory, to any morbid condition whatever that we have actually seen, and of which the common course and issue are known.

Take for example the opinions of Dr. Hughlings Jackson, already quoted upon this subject (p. 87). 'The great elaborateness of the movements in chorea' has led this author to the conclusion that the convolutions are the parts diseased. But if the character of the movements requires this assumption, what shall be said of their seat and variableness? With the supposition of diseased convolutions we are led to expect aphasia and mental disturbance, but not heart disorder. We have also to face the fact that very limited injury to certain regions of the cortex, whether irritative or destructive, will produce either spasm or paresis, but never chorea. In the list of cases which Dr. Ferrier has collected, there is not one which lends the slightest support to the belief that the movements characteristic of childish chorea are among the consequences of any brain lesion as yet described or produced. But the disparity does not stop with the *character* of the movements; it extends to the particular muscles implicated in

the two cases. In cortical lesions the leg and the arm are affected, or the arm alone. It is notorious that in chorea the two hands or the arms and shoulders are far more commonly affected than one side or one leg. (Append., Tables A and B.)

It thus appears that the pathology which really takes into account and satisfies all the phenomena of the disorder, does so in fact by eluding anatomical description altogether. It may borrow its phrases; it is not restricted to what they imply. The gist of the explanation lies always in some convenient expression we are bound to accept, but which is so little definite as to advance us nothing in actual knowledge. And in the last resort the trammels of anatomy are broken through, and the condition of the cerebral ganglia in chorea is described as one of 'delirium.'

II. It is this that I mean in saying that *the hypotheses which provide the best for the actual phenomena of chorea do not in fact present any distinct picture of a definite morbid change.* Take, for instance, the late Dr. Todd's description (p. 86), already quoted, which refers chorea to a 'debilitated state of the system, which, with the aid of a peculiar diathesis, produces a more or less enfeebled nutrition, leading, upon adequate cause, to a disturbance of the centre of emotion.'<sup>1</sup> Or take Dr. Handfield Jones's statement that the 'motor or emotional centres fall into a state of paresis.' Or take the several beliefs variously expressed of some 'toxic influence,' or of some 'eccentric irritation.' Assuming the relevancy of each of these descriptions, by how much are we the better for them? Except for Dr. Todd's statement, which centres in 'enfeebled nutrition' (which as a fact chorea does not exhibit), the rest are but translations of what we see and know. Whenever the voluntary muscles are beyond control, we may reasonably

<sup>1</sup> *Functional Nervous Disorders*, p. 361.

impute blame to the region which presides over movement. Whenever there is over-emotion, we may credibly ascribe it to a disturbance of the centre of emotion. Similarly, we may have 'toxic influence' and 'peripheral irritation,' and each of these in its own way may generate this same 'peculiar condition' which we call chorea. But observe that whenever these wholly metaphorical accounts of morbid influence and irritation can be brought fairly into the light, whenever they venture to assign some definite pathological state, such as anæmia or impaired nutrition, then at once they can be met and confuted.

But it may be said the lesions of chorea are real enough, but from their nature 'they are not susceptible of anatomical demonstration.'<sup>1</sup> The symptoms are transitory and paroxysmal, and death comes too late to find any trace of them. In this respect, it is urged, the disorder resembles epilepsy and other convulsive or spasmodic diseases, the nature of which must be investigated at the time and in the working. It must be answered that the actual physical conditions upon which the paroxysms of epilepsy or of nervous asthma immediately depend, if not known, are at least being hopefully sought for. Take the epileptic fit, for example. There are the materials to work with in the pallor of the face, the condition of the retinal vessels, the definite methods by means of which the fit may be produced or averted, and its frequent dependence upon direct mechanical pressure or the retention of particular elements of disintegration. Not only in disease but by experiment, by the drain of blood, by section of one of the lateral columns of the cord, or by irritation of the cerebral cortex, we can *obtain* epilepsy. Whatever the *modus operandi*, whether the brain be anæmic or hyperæmic, or first one and then the other, there is ample evidence of a profound vascular change

<sup>1</sup> Dr. Hughlings Jackson, *British Medical Journal*, Dec. 23, 1876.

of some sort. The symptoms and associations of epilepsy are definite and invariable. It has a recognised place in the category of diseases. Very different is the position of chorea. There is no known means of producing it artificially, and no known disease which (to say the least) is habitually associated with it; while as regards the condition of the cerebral circulation during the indefinite time that chorea drags on, we have proof, upon every test that can be applied, that there is no change whatever of the kind that we are in the habit of recognising as morbid.

And even when anatomical expressions are abandoned, and recourse is had to more flexible language; when it is said, for instance, that the excessive movements in chorea are due to the same agency as those of epilepsy, namely, to 'discharges of grey matter, which, except for great irritability from over-nutrition, is healthy,' the course is not made much clearer. The same explanation can hardly be made to serve both for chorea and epilepsy. The explosive discharge by means of involuntary spasm (which is epilepsy) is quite different from that continuous restlessness and feeble consensus of the higher muscles seen in chorea, which is without spasm, and not wholly involuntary; which never occurs in sleep or without consciousness, and indeed intelligence; which no manipulation can produce; which is almost unknown with certain races of mankind, and never seen in the lower animals.<sup>1</sup>

<sup>1</sup> Even could it be established, which it cannot, that choreic movement is a symptom of cerebral disease at all analogous to epileptiform spasm, we should have next to enquire whether the mode of onset and the sequel corresponded in the two cases. The fact is, that there is no such correspondence, but a marked contrast. This is enforced by the very same authority from whom I am now quoting—namely, Dr. Hughlings Jackson. 'Epileptiform seizures indicative of local brain disease begin in the thumb and index finger of one hand; the over-movement of chorea is in *all* the fingers, and generally of *both* hands. In the epileptiform seizure the lower part of the face is most affected; in chorea the upper part of the

III. It may be contended in the third place not merely that all the morbid conditions hitherto described fail to account for the phenomena of chorea, but further, that it is *difficult to conceive of any morbid condition whatever, anatomically demonstrable, which is at all calculated to produce it.* We have seen already by what course of reasoning the physical basis of chorea has been referred to the convolutions. But suppose we place it there, by what further supposition can any material defect of a tangible kind (more distinct and demonstrable, I mean, than 'mal-nutrition' or 'trophic disturbance') be assigned to this region of which it may reasonably be affirmed that it is calculated to provoke chorea? Look again at the facts, look especially at the points in which the chorea of every day differs from the chorea that is really wanted to satisfy anatomical theories. The brain cortex has by this time been injured and irritated by disease and experiment in a great variety of ways. It may be assumed that every kind of response of which it is capable has been elicited from it. It has been shown by the experiments of Dr. Ferrier and others that certain defined areas of the cerebral hemispheres have special endowments, insomuch that their stimulation is followed by particular movements or groups of movements.<sup>1</sup> Thus we get complex movements of the several limbs like those of climbing or swimming, or walking; extension movement of the hand and arm; the combined movement of head, eyes, and pupils, which gives the attitude of surprise,

face generally. In epilepsy aphasia is common, and especially with affection of the right side; in chorea there is never aphasia, and the speech defect is in no relation to the side affected. After epilepsy there is exaggerated knee jerk on the side that was convulsed; after chorea there is nothing of the sort.'—See Dr. H. Jackson on 'Epileptiform Convulsions from Cerebral Disease,' Medical section, International Medical Congress, 1881.

<sup>1</sup> See Ferrier, 'On the Localisation of Brain Injuries.'



and so on; each of these movements being, so to speak, under command of the operator, and producible always so long as the particular centre is structurally uninjured. Moreover, these several centres of movement being all near together, and all included in that area of the brain which is in relation through the corpus striatum with the motor tract, movements of these various parts are apt to be variously conjoined, or the one kind of movement to precede or follow the other, just as the stimulation of one centre gets communicated to the next. And from movements of a precise and definite kind which result from the earliest stimulation of a particular motor centre, we pass on to a corresponding spasm, and from that to a paresis, these several gradations corresponding with increased irritation and the production eventually of structural injury. It must be added that the earliest of these results, that of combined movement, is chiefly known to us in experiment, while the later consequences, spasm and paresis, are amply illustrated in cerebral disease.

Now, suppose some lesion or other of an irritative kind affecting the convolutions to be so situated as to concern some one or other, or some group of these motor centres. Upon the foregoing data, what consequence might be reasonably expected to happen? Surely that the earliest impression of such irritation should be seen in some definite muscular movement, or in a combination of movements corresponding with the proximity and relative position of the several motor centres, the leg and lower part of the face suffering with the arm, the head and neck with the eyes so as to give a particular expression, and so on. Or if the irritation were limited to one centre, we should expect (arguing from every irritation of the kind that we know of) at first extravagant movement strictly localised, presently spasm or tremor of increasing violence, and at last paralysis, just as the nerve deterioration proceeded.

How far do the facts of chorea correspond with such expectation? I have already alluded to the anomalous and irregular manner in which the several parts of the body are implicated, the most sensitive and educated muscles being the most liable to attack, and the hands suffering most of all; but what is more noteworthy still, is the character of the movements. Choreia does not consist in the frequent repetition of any one muscular act, or exaggeration of any particular bodily expression. Still less is it spasm. The muscles are restless, jerky, and indeliberate; they combine so badly that the intention of particular movements, whether of the tongue or the limbs, is not apparent, while, so far as the special muscles of expression are concerned, it is not the look of surprise that we see or of any other emotion, but mere restlessness and vacancy. Choreia is, in fact, not absolute but relative disorder. The movement which is proper to the child would be accounted choreia in the man or woman. The movement which is fitting and convenient for one limb or feature would be choreia in another. Nay more, the movement which we learn to accept from one individual we should call choreic in another. True, such examples as these fail to represent extreme choreia, but it is not extreme choreia that we at present want, but the common method of the disorder.

But there is another element of the affection much insisted on by those who are in search for its physical basis—namely, *ataxy*. Here, again, we have but to watch the thing itself to see at once how this word fails to apply to choreia in the same sense as to the progressive disease which depends upon degenerative changes. A single example will serve better than any general description to make this clear. I take the instance of a girl of twelve, shortly described in Case 6, Chap. XI. *Ataxy* is very marked in this patient, but it is in the arms and speech muscles, and not in the legs. When directed

to clap her hands together, she moves the two arms with a jerk, but in such manner as not only to fail but to conceal the existence of any definite purpose. And the more so, that there is no deliberate direction of the eyes to the place where the hands should meet. The movement is sudden and reckless and wholly abortive. So it is in the next attempt and the next. At last, however, by a sudden effort, as if resolving to anticipate the perverted working of more deliberate will, the act is performed to perfection, the palms meeting quite accurately, while with this unexpected success subsequent movements of a similar kind at once improve in accuracy.

It is the same with handwriting. When a choreic child has improved sufficiently to maintain some feeble and precarious grasp, the first attempt at writing some easy word begins with much purposeless flourishing of the pen point over the place, until at length it hits the paper and shapes a letter or two. Then follow further superfluous hoverings in the air, when again the pen alights and more is added. So the task progresses by snatches, and what the written word signifies is a number of detached motor acts, now more now less feeble, the intervals being surrendered to mere riot (see facsimile frontispiece).

Every one will admit the contrast between such movements and those of ataxia. The cautious and painful way in which an ataxic patient sets about clapping his hands, the careful manner in which, with the help of the eyes, the hands are moved towards one another, yet just fail to meet, or meet by a happy chance, not fully as in the other case, but with their sides touching, all this is as characteristic of the disease and as unlike chorea as are its progressive stages, implication of the organs of special sense, preference for the lower extremities, and limitation almost to adult males. Add to this (if more needs to be added) that the ill consensus of choreic muscles

varies continually with the mental state. In the case just mentioned, the confidence of success very quickly restored the limbs to their full use. Upon a mental disquiet, however (occasioned by an incautious comparison of her complaint, in the girl's hearing, with demoniacal possession), the ataxy returned temporarily, together with the full violence of the original disorder, and such extreme difficulty in mastication and deglutition that for some twenty-four hours she could hardly be got to swallow. All these symptoms, however, subsided in the course of a day or two with the quieting of the mind, and the girl eventually became absolutely steady.

I should be content almost to rest the whole case upon one such example as this, upon *any* example, indeed, in which the cause and course and termination and exact manner of chorea are faithfully and fully set down. If morbid anatomy will serve at one point, it will fail upon a complete review, and can only be maintained by having regard to exceptional examples or to certain isolated symptoms at a particular stage of their development.

## CHAPTER VIII.

## CHOREA REGARDED AS FUNCTIONAL DISORDER.

The parts selected by chorea—Its varieties and modifications—Character of its ataxy—Its form in relation to its cause—Alliance to hysteria and emotion—Manner of progress and of recurrence—Objections.

MORBID anatomy thus failing to account for chorea, we are led to consider the question of its pathology from a different point of view. May it not be looked upon as a functional disorder, a motor disturbance which is as much beyond the reach of anatomical demonstration as are the various passions and emotions which, like it, distort and coerce the body in a great variety of ways? This functional hypothesis will, no doubt, be rejected by some as a mere evasion of the difficulty, and at the outset I attempted some answer to that preliminary objection. Assuming, however, that we may properly recognise certain defects in the exercise of the muscular functions apart from material disease, the question still remains whether or not the phenomena of chorea, or any of them, may properly be reckoned amongst these defects. We have seen that anatomical theories are faulty and inadequate. How far can any functional hypothesis take their place? It will need to be tested like the rest in the light of the facts, by considering, that is to say, the character of choreic movement, its subjects, causes, and associations.

It has been urged in the preceding chapter that the movements of chorea, in their method and mode of distribution over the body, together with the rarity of spasm and the

transient character of paralysis, give no warrant whatever for assuming the existence of any definite morbid change, while, as a matter of fact, no such change has been found. In their variety and half voluntariness such movements would seem, as we have said, to have their source in the cerebral convolutions; yet not in convolutions diseased in any of the ways which anatomy can discern. *The limbs become subject to chorea in the order of their use as intelligent instruments*, and not as they would do upon any assumption of injury of a motor centre. Thus, untaught muscles, and such as have never been employed as the agents of intelligence or of some definite emotion do not suffer chorea; while, further, the more complex the muscular employment, the higher the place in the intellectual scale of any particular limb or group of muscles, the greater the liability in this respect. The arms suffer chorea much more than the legs, the hands much more than the arms, the right hand more than the left, and the face (so soon as sensitiveness begins to settle there) more than any other part of the body.

Chorea thus indicates the withdrawal or infirmity of controlling power over certain parts of the frame, and will be seen, *cæteris paribus*, first and most in those muscles which are by nature and the process of education the most unruly. Its precise manner, at whatever age it may occur, is due to this: that the muscles concerned, having been trained more or less completely in certain kinds of movement, are suddenly thrown back in their education by some nervous shock, which renders them restless and unruly, while the operation of the will no longer sufficing to regulate the limbs, tends rather to disfigure the movements it is unable to arrest. Yet the child thus disturbed does not revert to the method of infantile movement. There still lingers some memory of order, and the simpler uses of the muscles in their acquired conjunction are in a measure retained.

But the capacity for stillness, which ought to develop *pari passu* with the acquisition of purposive movement, is enfeebled. Intelligent movement is for the while embarrassed, disfigured, and overborne. It is not lost, and under favouring circumstances, or by strong effort, it will assert itself. A sudden access of determination or confidence, a purely mental operation, will bring the limbs for a moment to order and quiet. But the habitual condition is one of weak restlessness when there should be quiet, and of exaggerated misdirected movement when some muscular act is attempted.

Hence it is that the deformity of the affection appears more and more as life goes on. In early childhood the over-movement is easily concurred in and hardly deforming, but in adult life, the will being in active opposition, the disorder exhibits a corresponding distortion.<sup>1</sup>

We are thus led to consider chorea as being no definite thing susceptible of one general description like spasm, or tremor, or paresis, or any other of the clinical signs of nervous disease. It varies with its subjects. The child who has not yet acquired the full use of its voluntary muscles, whose movements are undecided and featureless, is the proper and, indeed, the only subject of that irregular and variable commotion which the books seek to describe. The mismovement is but an exaggeration of the natural movement at that time of life; a mere restlessness which is without character or rhythm. But when chorea comes to the adult, his fixed habits of moving will impart to the disorder more distinct method; the affected

<sup>1</sup> Apart from chorea this same law is of universal application. Thus when, as often happens, the face muscles pass beyond control so as to express sentiments which it is desired to conceal, there results from this conflict a localised chorea, a chorea which has many degrees, from the slight facial disorder which indicates that some restraint is being put upon the features to the absolute distortion of face, which occurs when some violent passion is to be concealed.

limbs, that is to say, are jerked about after a manner that may be described; the mismovement is more or less rhythmical; or there is a recurrent twitching of certain muscles in a constant order. And as life goes on, chorea presents these characters more and more, insomuch that we might guess with some accuracy the age of a choreic person without the aid of the features.

And along with these several modifications of chorea, regarded as a motor disorder, there are others which concern its mental or psychological associations. In these respects it not only varies with the age and temperament, it is affected visibly by passing events, and may be of one form to-day and another to-morrow. In any moderate chorea, as was mentioned in its definition, if the attention of the child be expressly called to a particular limb, and she be bidden either to increase or to modify in any stated way the movement of that part of the body, the effect of such observation will always be to alter for a while the character of the restlessness. Similarly, the holding of one limb will have the effect of transferring the over-movement to other parts of the body;<sup>1</sup> the restraint is never actively resisted.

The mental or psychological connection is seen still further, as was shown in the last chapter, in the *ataxy* of the affection. It is to be remembered, indeed, so far as anatomy is concerned,

<sup>1</sup> Some years ago a country youth was under my care, a farm labourer of good intelligence and robust frame, who, when answering questions, had an agitation of the right arm and shoulder, coincident with speech. There was no want of co-ordination in the limb, and no impairment of use. When the agitated arm was forcibly held by one of us during his talking, the agitation was transferred to the arm and shoulder of the other side. The same transference would occur when the boy had occasion to speak while the right hand and arm were employed, *e.g.*, when holding a pail of water. And not only in these ways, but by directing the patient to keep his left (or unaffected) arm in constant motion while speaking, the usual agitation of the right side would be arrested.



that morbid changes in the cerebral cortex do not provide for ataxy at all. But what may be noticed here is that the ataxy of chorea, like its over-movement, is of that kind of which we get the pattern—I do not say the degree—not in structural disease, whether cerebral or spinal, but in the many conditions of mental excitement and perplexity to which all mankind are liable. A choreic child may fail utterly in some muscular act one day which she will accomplish easily the next. The very same purpose which cannot be accomplished when set about deliberately will be performed accurately on a sudden, and without premeditation. Again, observation by others has the effect sometimes of aggravating and sometimes of controlling choreic ataxy; or it may have both these effects at different times in the same subject.

It has been urged by Dr. Gowers that the ‘inco-ordination of chorea is distinct from mere inability of the will to still the muscles,’ and he thence argues that the independent variation depends on a morbid condition of distinct and perhaps separate regions. But it is to be remembered that in nature muscular co-ordination and the power of muscular stillness do not commonly vary together. There are many habitually restless individuals who will at once bring their muscles into order when employed on some familiar task. There are others, still and reposeful so long as they do nothing, yet painfully wanting in co-ordination when set to work. In so far, therefore, as chorea may properly be said to depend upon a mere exaggeration of defects which all exhibit in some degree or other, the manner of its ataxy, as contrasted with its restlessness, is just what we might expect.

A further argument in favour of the functional hypothesis may be drawn from the observation of the actual starting place of chorea.<sup>1</sup> In young children, the hands (one or both,

<sup>1</sup> See Appendix. (Tables A and B.)

*but the right much more than the left*) are in a large number of instances the parts first disturbed. Now it is obvious that in childhood there is no member of the body more heavily taxed than this. In the modern requirements of education, the right hand of the school child is habitually overtaxed. And accordingly, without obvious nervous shock or previous rheumatism, we get hand chorea in school children, the particular cause of which is regarded as very obscure. Such examples, however, often receive their explanation, if we accept the doctrine that the work and competition of school may furnish occasion for mental disturbance which finds expression in the part most exercised.

Other examples may be quoted which tend to show that, along with the general law which makes the muscles of the highest endowment the most liable to chorea, there is a tendency on the part of those of them which are the most directly beset or embarrassed to suffer the disorder first and most. The operation of this principle is seen generally in the successive modifications which chorea undergoes at the several periods of life, the limbs often suffering alone in childhood as being the instruments of emotion ; while at a later stage, when the face comes to be a conscious index of the mind, it is the facial muscles which are the most disturbed. It is seen also, individually, when the particular event or catastrophe which occasioned the affection happens to concern a particular group of muscles. A boy of eleven was much alarmed by an explosion of gunpowder, which slightly scorched his face, and was followed by choreic twitching of the eyelids. A girl of eight and another of ten, having accidentally pinched the fingers of one hand, became choreic in those fingers. A boy of eight gets chorea of the hand in which he was holding the reins of a runaway horse. A girl of eleven, stooping to pick up, as she thought, her brother's cap, found to her great

alarm that she had seized a dead rat: she became choreic in the offended hand (the right), and so markedly that, although the general chorea which followed was not severe, this one member for a long time after could by no effort be kept still.

Similarly I have noticed repeatedly chorea originating on the side whence the alarm comes. A boy frightened by a dog jumping up at him gets chorea on the side of the dog's approach; a girl frightened by a man suddenly starting out upon her in the dark gets chorea of the side exposed, and so forth.<sup>1</sup> A very large number of cases would of course be needed to verify the hypothesis which these examples would appear to illustrate; but I think the number might be found, while the principle itself is no other than that of which we have familiar knowledge, in the after agitation of muscles which have been strongly excited, and their tendency to recall and even repeat the violent movements of emotion or passion.

And observe that in the most violent chorea the muscles still maintain their accustomed concert. The muscular over-movement, except for its exaggeration and disregard of the will, is the same as the natural movement. Putting aside the paresis which in varying degree is conjoined with chorea, it is not true to say (as has been said) that it will select certain muscles out of a group, and move them after a manner of which mere will is incapable. That this conjoint action of the muscles remains intact is shown especially in this, that in

<sup>1</sup> The above are cases of my own. I could quote many more to like effect. Ziemssen relates a case of violent chorea of the legs reaching great height in a few hours, occurring in a boy, owing to fright, from a gun being fired behind him when he was gathering hops.—'Cycl.,' vol. xiv., p. 432.

It is to be observed of these instances and of others like them, where chorea is caused by some grave terror, that, contrary to what may be regarded as the rule of the disease, there is hardly an interval between cause and effect.

those parts of the body where the several muscular uses are most distinctly differentiated, there, in exact proportion to the degree and perfection of such differentiation, do we find a more or less perfect separateness of choreic movement. Thus, for example, the hand and the fingers are concerned in many tasks in which the arm takes no direct share. And accordingly the hand and the fingers are often choreic without the arm. It is the same in regard to the two sides of the body. Movement of one arm is common enough in ordinary life, without need or suggestion of movement in the other; but the movement of one leg almost implies movement of the other. Accordingly, chorea of one arm alone is very common, and chorea of one leg alone is very rare.

But we have yet to consider the disorder in its connection with mental disturbance, hysteria, delirium, and mania. Zeimssen, indeed, is content to refer 'the deterioration of intellect to slight anatomical changes in the central apparatus of the nervous system.' But such an explanation, indefinite as it is, will not serve. Deterioration of intellect is by no means common. While many patients with severe chorea escape cerebral symptoms altogether, the liability to mental disturbance is governed most by the age; it varies independently of the motor disorder and is of all degrees, from slight emotional display to acute mania. Any account, then, of the mental phenomena which shall connect these invariably with the over-movement is contrary to the facts. What has to be explained is this: that chorea in the child is usually a purely motor affection; that mental disturbance comes to mix with it at and after the time of puberty, and that the two symptoms sometimes combine and even alternate.

This gradual intrusion of the emotional element, the transmutation, so to speak, wholly or partially with growth, of a motor into a mental disorder, is a phenomenon which

finds ample illustration in other affections, and especially in hysteria. In the hysteric fit there is often a sudden and complete transition from the sighs and tears which first announce it to the violent convulsion which at once dissipates it, until again exchanged for emotion. At one time the hysteric fit will be purely motor, a convulsion or relaxation of the whole frame; at another it will be purely emotional, a burst of laughter or of weeping, or of these two combined.<sup>1</sup>

But the manner in which the voluntary muscles are put to use, both to express and take the place of emotion, is best seen in childhood. Childish grief is actually dispelled by means of movement, and conversely any violent bodily exercise will always bring children to the verge of emotion. A little child in sudden affliction, reserving as long as possible the motor expression of it, will suffer the while in mental respects; presently a slight quivering of the body, with catching of the breath, and irregular or perhaps suspended respiration, indicate the approach of relief, and this will come at last in the convulsion called sobbing, soon to be followed by perfect mental serenity. In such familiar facts of our nature, we find the real clue to the various ways in which the mental phenomena of chorea associate themselves with its motor disorder at different periods of life.

From considerations such as these, suggested by the method and movements of chorea and the parts it chiefly affects, we may pass on to those which concern its progress

<sup>1</sup> In later life tears and laughter and spasm no longer come readily as convenient avenues for feeling. Yet there is still a motor equivalent, or exchange for emotion. It consists not in over-movement or spasm, or any of the violent muscular displays which are associated with early life, but in some slight over-tension or relaxation of certain muscles. After mental shock or affliction in later life comes a less erect posture of the body, an immobility of the face, or, it may be, an alteration of the whole carriage.

and development. Chorea is usually separated from its exciting cause by a distinct interval. In many instances, however, this interval is more apparent than real, and arises from the fact that the motor part of the disorder develops very gradually, and fails at the first to attract notice. Yet so soon as it is once recognised, whether by its subject or by those about him, it rises at once into prominence. From this point its progress and duration are extremely variable. Sometimes the patient's first consciousness of infirmity coincides with the height of the disorder, and after a short interval when both the disease and its treatment have become familiar, the over-movement begins to decline. In some severe cases with the elder patients it will often thus moderate with remarkable rapidity. With very young children, however, the course of chorea from the time of its first recognition is one of continual ups and downs, until at last and it may be quite suddenly an unusual improvement is noticed, from which time the progress to recovery proceeds rapidly and continuously. Yet even now this favourable progress may at any time be arrested by a mental shock, and the patient thrown back to his worst state, or it may be to worse than that.<sup>1</sup> This uneven progress, with the striking manner in which the first distinct sign of amendment becomes the foothold for further recovery, is among the many characteristics of chorea testifying to its functional basis. Not less pertinent towards the same end is the long-enduring chorea of young children, where the patients are both indifferent to their disorder and incapable of applying the effort or attention necessary for its cure.

But we are to notice further that as life progresses the time presently comes when we are finally secured against chorea. As with experience and use the voluntary muscles acquire

<sup>1</sup> See, for instance, Cases 6, 11 and 13, Chap. XI.

variety, so do they acquire stability, and thereupon lose the infirmity of childhood. They become less and less subject to chorea, not because they have escaped any of those liabilities to embolism and nervous deterioration which are said to produce it, for these liabilities are every year on the increase, but because use and variety of employment promote muscular accuracy and obedience. Thus there is nothing which so certainly removes the individual from the influence of chorea as physical training and education. The more nicely adjusted are the several uses of the higher muscles, or, if the phrase be preferred, the more distinctly differentiated the functions of the cortical centres, the greater is the power of control.

And not merely does age with its fixed uses deliver the body from the assaults of chorea, but further, the several departments of the muscular system escape from it one by one in the order of their education. Thus, for example, the higher employment of the fingers in writing, sewing, and the like, are not self-taught; they are the result of laborious training and exercise. There is nothing in the natural wants of childhood to suggest the way to them or to recommend their employment. Yet after a while the time comes when these intricate movements are performed as readily and automatically as any. It is only then that they become as fixed and unvarying as the rest of the muscular habits. I write on this page with as little thought, except at this instant, of the complicated shapes my fingers are forming, as I shall presently give to the movements of the legs in walking down Bond Street. So soon as the hand and fingers are thus brought under the very same subjection as the lower limbs, they are protected, or in large measure protected, against chorea.

And what is true of the hand is true also after its manner of other parts. So long as the tongue or the face or the speech muscles are being taught and gathering experience, so long do

children hold precarious possession of those parts of their bodies. But when long use and constant repetition have made the higher bodily movements, each in its turn, as natural and spontaneous as the movements of the legs in walking, we begin to hold them with something like the same security. And at last with the completion of the muscular education, a stage which is never reached except in company with intelligence and mental stability, true chorea, after the child's pattern becomes altogether impossible, be the pathological conditions what they may. Overstrain, fatigue, and other influences directed against the voluntary muscles do not cease to be injurious, but the nature of the response is different. Just as the overworked hand of the school-girl suffers chorea, so the over-worked hand of the law-writer suffers palsy or trembling.

Again, what hypothesis except the functional can be made to provide for a disorder which is recurrent as chorea is, recovering perfectly time after time; and at last, when adult life is reached, finally disappearing, the signal for this departure being nothing else in the world but the departure of childish ways and the substitution of new modes of response to nervous impressions? Observe, too, that during the time that this liability to recurrence continues, it is not the same muscles that are implicated in successive attacks, but rather those which are similar when regarded as intellectual instruments. The manner of the repetition does not accord with any notion of altered nerve-centres influencing a particular group of muscles again and again; it accords with the more assured observation that emotional expression makes use of different parts at different ages. Thus the recurrence of a childish chorea in adult life will not necessarily be an exact repetition of it, although in obedience to exactly the same law. The muscles which at the time are the most sensitive are still the muscles which suffer. The generalized chorea of early childhood when repeated later



on will occupy chiefly the hands and the face because in the interval the individual has become self-conscious. A further principle is involved in the case of chorea happening repeatedly to the same side, if not to the same limb. The fact may seem at first sight to need some anatomical explanation, but no one will deny that muscular failures and inco-ordination are long retained and easily recalled. In the articulation of certain words and the performance of certain manual and facial acts the same infirmity of a particular group of muscles will appear again and again, however long the interval between one attempt and the next. To say that the muscles remember, and that the same muscles are repeatedly disturbed owing to the repetition of an old impression is, it is true, only to say that the first impression has wrought some change or other in the brain, but it is a change to which no material conception attaches.

To these arguments, drawn from the nature of the choreic disorder, its variations with age and with mental development, as well as the manner in which it originates, progresses and recurs, others may be added having reference to its exciting cause and favourite subjects. Of the immediate causes of chorea there is none than can be put into comparison with the mental. And not only in actual frequency does it stand first, it is superior to all the others in the intimacy of its association with the disease it occasions. We are invited to accept rheumatism as the cause of chorea, whatever the interval separating the two affections; and in fact, as has been shown, they are seldom actually consecutive. But in the case of fright there is often no interval whatever, while many other forms of nervous disturbance, such as the afflictions of schooling, though slower in operation, have chorea as their direct consequence. And when puberty is reached the immediate associations of chorea need the same interpretation as its childish symptoms. There is no condition so

intimately connected with it as hysteria.<sup>1</sup> The two disorders, although properly separate, have so much in common that the very same symptoms which we call chorea in the child we call hysteria in the woman, while not seldom motor and emotional infirmity get so combined that we see in the same subject now one and now the other affection the more prominent.

For such reasons as these I regard chorea as mainly a disorder of function. Some structural basis it has no doubt, like all other movement, orderly and disorderly, and its remarkable connection with endocarditis is not to be forgotten. Still, in its ordinary dress it rarely displays symptoms which can be traced to any material morbid process whatever. The disorder most nearly allied to it is one which, like it, is transient and recoverable, and attaches itself to the same sex at a somewhat later period. To explain chorea we must look beyond it, and especially we must look to that multiform disorder whose physical basis has yet to be sought, and which at the next epoch of life, when in its turn the emotional part becomes unduly prominent, is apt to arise in the same sex, and upon similar incentive in strict parallelism with this motor disorder of children.

<sup>1</sup> It is interesting to notice the attitude of pathological anatomy towards hysteria as contrasted with chorea. The following is from Jolly's article on Hysteria in vol. xiv. of Ziemssen's 'Cyclopædia,' p. 488:—'That some alteration in the nature of the nervous system must be at the foundation of its altered function cannot be doubted. But whether we are to expect that these changes affect the structure of the nerve elements, or only have reference to its chemical and molecular constitution, cannot yet be stated. It is quite as possible that the microscope may at some time give us information concerning the nature of this condition, as it is that no information may be derived from this source. One can, therefore, in the meanwhile only discover the basis of hysteria in an "abnormal" condition of the nervous system, which can only be determined by its symptoms.' Thus coupling this passage with what has gone before, it appears that pathological anatomy gives us a "delirium of the corpus striatum" to account for chorea, and "an abnormal condition of the whole nervous system" to account for hysteria.

Against these assertions certain objections are raised : some of them have been already mentioned and some may be briefly noticed in this place. They may be classed thus :—

1. Chorea is an enduring affection which often does not even commence until its supposed cause has ceased to operate.

2. Chorea occurs in connection with paralysis, convulsion, and other similar phenomena indicative of cerebral or nervous lesions, if not uniformly, yet too often to admit of the supposition that the connection is accidental ; while, added to this, chorea ending in death actually exhibits definite morbid changes, endocarditis as a very general rule ; and nerve changes, such as minute emboli and softening, exceptionally it may be, yet in an aggregate too large to be neglected.

3. Chorea is commonly expressed in the limbs of one side more than the other, and exceptionally it is wholly one-sided.

1. As regards the *duration* of chorea the facts are these. It is apt to be much lengthened out in young children, is of somewhat shorter duration in those that are older, while at the puberty age, or when connected with pregnancy, it is severe but not long. In later life and in old age it is a permanent, incurable disorder.

Now, if the account I have given of the circumstances which attend chorea be a true one, these several modes of it are not unaccountable. In childhood, as we have seen, the muscular system, disturbed by some nervous shock, degrades from the orderly movement which education was developing towards the aimless irrepressible movement of infancy. And if this be so, what is the probability of an immediate restoration ? From what source is that extra power of control to come which will enable the child suddenly to quell such a disorder as this ? From nothing that I can think of except that gradual strengthening of the will which comes naturally with development. Children do not immediately recover from

chorea ; young children, unable or unwilling to make an effort on their own behalf, retain the disorder for a considerable time, yet they all recover eventually, while the circumstances promoting or retarding their recovery clearly indicate its mental dependence. The growing power of the will together with the natural decline of childish restlessness are pledges for ultimate relief.

In the chorea of grown-up girls and young women the conditions are somewhat different. There is a disturbance of mind as well as of body. Both are on the side of disorder. The muscular agitation provokes and maintains a corresponding emotional excitement. Yet such chorea, although severe, is not often prolonged. Excited sometimes by pregnancy, and sometimes by the arrival of the catamenial period, it also, like the other, has security for its ultimate cure in the temporary character of the conditions to which it owes its origin. The chorea of the puerperal state, indeed, as Dr. Robert Barnes points out (p. 34), has often for its subjects women who have already suffered in the same way in childhood, and in whom the occurrence of pregnancy so arouses the emotional element as to render their power of control once more, for the while, inadequate.

It is otherwise when we come to consider chorea in adult life and in old age. But the same reasoning applies to these as to the others. Choreia may persist with the advance of years because the childish mind persists, and that extra will-command which we expect with the natural development, and to which we trust always as a curative agent, never comes. Or otherwise chorea may happen when life is already advanced, but only to those who are intellectually weak, or who from age have come to share the infirmities of childhood. But come as it may, it is obvious that in adult life the disorder has not the same hope of recovery as at an earlier period.

There is no reinforcement of will power to be expected. The disorder has gained ascendancy over the individual at his best. There is nothing in the man's future, as there is with the child, to dispossess it.

2. But it is said *chorea occurs in connection with hemiplegia and convulsions*, symptoms which appear to depend (and indeed in fatal cases are commonly found to depend) upon definite structural lesions. The statement must be admitted, but it is incomplete. We have to add that chorea occurs much more often without these accompaniments than with them. And further that the circumstances of these exceptional cases forbid our accepting them as illustrations of the pathology of the affection *per se*. Chorea as a rule occurs by itself. In its common form it is a temporary affection of childhood which neither wastes the muscles nor paralyses them, except temporarily, and which disappears when childish habits have subsided, leaving no trace behind. But we need not stop here. Let us consider the actual significance of those cases in which chorea and paralysis are so associated as to appear due to a common cause.<sup>1</sup> All such examples exhibit not chorea merely, but chorea disfigured owing to this very combination.<sup>2</sup> We are, in fact, in this position, that so long as the affection exists alone it furnishes of itself no sufficient ground for asserting any such morbid change as anatomy would recognise; while so soon as other symptoms are superadded, these so complicate the matter as to make the proper *post-mortem* signs of the simple affection extremely difficult of separate recognition.<sup>3</sup>

<sup>1</sup> See, for instance, Case 16, Chap. XI., with the remarks there appended.

<sup>2</sup> There remains, indeed, the question of endocarditis. This is discussed in Chapter IV., and will be again referred to under Pathogenesis.

<sup>3</sup> See, for example, Dr. Tuckwell's cases, *Med.-Chir. Review*, Oct. 1867, and *Bartholomew's Reports*, vol. v.

Brain lesions of whatever kind do not, as a rule, exhibit chorea at any part of their course, and conversely, chorea implies no appreciable liability to cerebral disease. Nay more, cerebral symptoms, such as vomiting and partial convulsion, so readily excited in young children, are but seldom seen in this connection. All the morbid conditions of the nerve centres that have been recognised are much more often without chorea than with it, yet *any* disturbance, even such as is not directly cerebral, may revive or even produce it.

3. It is further objected that *chorea commonly affects one side of the body more than the other*. Whatever the force of this objection to the functional hypothesis, it must in fairness be added that chorea is but rarely confined to one side, and that it sometimes changes from one side to the other. (Case 3.) It is true that the arm will commonly be affected along with the leg of the same side, but the manner of the attack is quite different from that due to actual brain lesion. Thus the hand or less often the foot will be first affected and next both arm and leg will come to share the same disorder. But it is quite as common to find that after the hand has been affected, the disorder will pass thence to a limb of the other side, while the commonest mode of beginning of all (excepting perhaps the face) is in the two hands. (Appendix, Tables A and B.)

That the foot should share the disorder of its own side when both it and hand are affected will not appear wonderful when we consider that limb movement of whatever kind, as a mental expression, follows the same rule. The fact is but little apparent owing to the separate employment of the upper and the lower limbs. The legs are for the support of the body and for progression; being wanted for standing and walking they are not available, and in adult life are seldom made use of, for emotional

expression. But children, as well as savages, express themselves with their legs, dancing, jumping, and kicking in many moods. Moreover, making allowance for the maintenance of balance, the leg is always apt to sympathise with the arm of the same side. In dancing, for instance, as the arm is flung up the foot of the same side is thrown out, and in beating time to music, an action not confined to children, so soon as the measure becomes lively enough the foot will be stirred in sympathy with the hand and arm of its own side. Further, the occurrence of emotional hemiplegia may be mentioned, a very temporary paralysis, due to excitement, which is quickly and completely recovered from.

But while we properly insist upon keeping the pattern of simple chorea distinct and separate, there is reason to believe that in some rare instances the long continuance of motor disturbance may ultimately give rise to structural deterioration of the centre on which such movement depends. It is difficult without some such hypothesis to account for chorea in some rare instances becoming permanent, for the occurrence of hemiplegia as its immediate sequel, or for limb distortion owing to contraction of the flexor muscles (Case 17). The pathology of such cases is confessedly obscure, and until more light is given it can serve no purpose to speculate about them. Yet the more we lay stress upon the very few examples having such issues as these the more we bring into prominence that general law of the disorder which exhibits it as a temporary affection wanting in all those symptoms which imply nervous structure change. Chorea, in a word, for its full development requires the service of a brain and cord structurally uninjured, and so soon as textural injury intrudes it will alter its proper form, and give room to some one or other of the recognised symptoms of nervous disease.

It remains to consider how such conclusions as these may

best find expression. There is an ample vocabulary now at the disposal of everybody, which has been designed to cover many ill-understood nervous conditions. Impaired nutrition of nerve-centres; disruption of the normal relations between such centres; excessive, defective, or misdirected nervous discharge by cells of some particular part of the brain cortex, these and similar expressions, commit us to nothing, and in so far as they have a precise meaning, may find ample justification some day. Meanwhile we may legitimately direct attention to the near alliance of chorea with those emotional disturbances of which both the origin and the remedy are universally known.



## CHAPTER IX.

## PATHOGENESIS.

Pathological relations of arthralgia and of endocarditis—Symptoms common to chorea and rheumatism at different ages—The early kindred and later separateness of the two affections.

IN the preceding chapters certain facts have been adduced in favour of considering chorea as essentially a functional disturbance. It has been shown at the same time that, inasmuch as the affection is associated with rheumatism and with endocarditis, no purely functional hypothesis can suffice, and reasons have been given for regarding chorea as of the same pathogenesis with acute rheumatism, the two diseases being virtually distinct, though owning a common origin. A few concluding words may be here added as supplementing what has been already said on this subject (p. 76).

Facts are accumulating which render it probable that the common symptom uniting a group of nervous phenomena is arthritis or arthralgia. To narrow this hypothesis by the use of any epithet, such as rheumatic, and so to lump together the joint pains of chill, of growth, of fatigue, of many ill-defined neuroses such as hysteria, and it may be added of chorea itself, is misleading and gratuitous. 'Any form of nerve lesion,' says Dr. Weir Mitchell, 'the brain included, may develop in the joints inflammatory conditions, usually subacute, which so precisely resemble rheumatic arthritis that no clinical skill can distinguish them.' One illustration of this is the occurrence of polyarthritis indistinguishable from the rheumatic

in recovering paralysis.<sup>1</sup> Similarly, joint pains that commonly pass for rheumatism may arise from nervous strain. It is a perfectly unwarranted assertion that chilling of the body is the sole cause, or even a necessary factor, in the production of so-called rheumatic arthritis. Years ago Corrigan called attention to the fact that fatigue has its share in the production of rheumatism. More recently Senator<sup>2</sup> suggested that mental emotion might contribute to produce the disease, quoting an instance of the sort in his own experience. It is not uncommon to meet with children suffering polyarthritis, so-called rheumatism, where there is a history of fatigue but none whatever of exposure. 'There is a growing belief,' says Dr. Weir Mitchell, 'that rheumatism may be of more forms than one.' It would, perhaps, be safer to say that there are many other forms of joint pain besides the rheumatic. Amongst these may be instanced that form of it coming and going with absolute suddenness, of which we have examples in chorea and hysterical knee.<sup>3</sup>

But while there is no want of illustrations of a near sympathy between nervous disturbance and arthralgia, speculations

<sup>1</sup> Sir William Gull published (*Guy's Hospital Report*, 3rd series, vol. iv.) a suggestive case of temporary paralysis, following severe and unusual exertion in a woman of 39, where the larger joints were affected in a manner indistinguishable from the rheumatic. Many similar cases have since been recorded.

<sup>2</sup> Ziemssen's 'Cyclopædia,' vol. xvi.

<sup>3</sup> In a recent lecture (*Philadelphia Medical News*, April 22nd, 1892), Dr. Weir Mitchell quotes some cases of post hemiplegic joint pains occurring within four days of the hemiplegia, and others where they have preceded it. He points out that this rapid development is inconsistent with any theory of descending degenerative change involving the motor tract and cord. 'Really,' he says, 'it does not seem quite certain that the joint trouble, as well as the pain, may not have their origin in the cerebral centres.' And further of the acute unilateral pain which precedes hemiplegia he says, 'I have myself no doubt that pain and many other sensations may be of cortical origin.'

as to the nature of such sympathy are speculations merely.<sup>1</sup> It is enough to say that the joint pains of chorea are fully provided for without any reference whatever to rheumatism.

Precisely the same may be said of endocarditis. It has been shown that both anatomically and in its clinical course the endocarditis of chorea is apart from that of rheumatism. (Chapter IV., p. 61.)

But although neither joint pain nor cardiac murmur is any certain indication of rheumatic kindred, yet it is inevitable that the occurrence of these two symptoms in chorea as well as in rheumatism should suggest a certain likeness between the two affections, and this the more in the case of children whose arthritic symptoms are often equivocal.

And there is this further and independent fact that in early childhood, but at no other period of life, undoubted rheumatism is seen in *frequent and intimate connection* with chorea, and that this connection, loosening with years, becomes hardly perceptible in later adult life (p. 49).

This relationship, thus varying with the years, will appear the more plainly by placing side by side the two affections, chorea and rheumatism, and observing their attitude towards each other from early childhood to full age. When first the two emerge in recognisable shape they are seen in close companionship. A little later, say in middle childhood, chorea has attained its full proportions and exhibits both over-movement and cardiac disorder, it has also reached the period of its greatest frequency. Rheumatism, on the other hand, remains still ill-

<sup>1</sup> The occurrence of gastric crises, together with nerve, joint and bone affections in locomotor ataxy, has suggested to Dr. Buzzard ('Dis. of Nervous System,' p. 266, &c.) the hypothesis of 'a trophic centre for the osseous and articulatory system in immediate neighbourhood of the roots of the vagi.' Whatever may be said of this theory, awaiting its physiological proof, it is not greatly furthered by chorea, where, notwithstanding that the subjects are children, gastric disturbance is conspicuous by its absence.

defined and difficult to identify. Nor is it until near puberty that acute rheumatism acquires its characteristic development with marked pyrexia, swollen and tender joints, profuse acid sweating, and such general stiffening of the limbs as tells its tale at a glance. Meanwhile, and side by side with this clearer definition of rheumatic arthritis, the chorea of the same period has become less distinct than at first. And it is less frequent as well as less express. No longer a purely movement disorder, in which the heart is affected in a remarkable way, it has become emotional as well, and more or less nearly allied to hysteria, while the endocarditis, which in childhood it shared with rheumatism, is now absent. Thenceforward in the progress of life the pattern of acute rheumatism is maintained for some years; but chorea ceases to occur, or in its rare appearance is so altered from its childish form as to be hardly recognisable.

If, reversing this order, we work up the stream of time from the adult to the child, we have at the former period these two affections showing no sort of alliance. The one exhibits emotional over-movement without paresis, and without heart disorder (the form chorea takes in young women); the other shows the distinctive features of acute rheumatism, in combination often with endocarditis. But continually, as childhood is neared, these two separate affections approach each other, and the most distinctive period of chorea (the proper chorea, as we may say, of childhood) is also that where it displays two remarkable symptoms, endocarditis and joint pains, both of which it shares with acute rheumatism.

Thus the evidence of a common birth is afforded not only by the frequent concurrence of these two diseases in early life (which is matter of demonstration), but also in the community of their symptoms at that period. With growth they get differentiated, and the interval that separates them goes on widening with the years.

And if the nature of rheumatism and of chorea be considered and the tendency of both to recur, the reason for this differentiation will appear. At first in early childhood when their affinity is the most conspicuous these affections will alternate or even concur. But there are causes productive of chorea and causes productive of rheumatism, and so the changes and chances of life will favour development in the one direction or in the other, while every repetition of either affection will strengthen its own hold and weaken that of the other. One illustration, taken from an earlier part of this book, may make my meaning plainer. A girl of three years and six months (p. 50), among the youngest examples of this kind on record, showed rheumatism and chorea intimately mixed, a group of symptoms motor, cardiac and articular, signals in common of a nervous disturbance of which the cause was given. Here were the elements out of which two distinct affections might have been constructed. But at the time they were not two but one, now the rheumatic, now the choreic element preponderating. And by their joint appearance at this exceptionally early age they indicated a special liability both to chorea and to rheumatic arthritis. Supposing this child to have lived, it is probable, upon the analogy of similar cases, that she would in her after life have developed either chorea or rheumatism; or now one and now the other, according to circumstances, exposure on the one hand, nervous shock on the other. But this comparative indifference of response would be of short duration. Whichever disorder she encountered first would impress its own likeness and tend to weaken the birth-born alliance.

Speaking of chorea generally there can be little doubt that its relation to rheumatism is often exaggerated, while the extreme opinion expressed by a few that the one affection is but an expression of the other is demonstrably wrong. But a

consideration of the question from the point of view of age compels the conclusion that chorea and polyarthritis are in their pathogenesis nearly akin ; it furnishes the reason why these two affections, though cradling together, should have but a short-lived intimacy, and suggests the agency which soon begins to relax the bond of union and eventually altogether dissolves it. Both chorea and rheumatism are, it is probable, members of a pathological group which has arthritis for a common factor, and for whose underlying source we are yet in search.

## CHAPTER X.

## TREATMENT.

Early management—Feeding—Procuring sleep—Encouragement and allowance—Lingering—'Acute' chorea—Prostration—Relapse—Question of drugs—Convalescence—Summary of treatment.

EXCEPTING insanity, there is probably nothing more disturbing to home life than chorea, and the older the subject the greater the scare. It is not the patient alone that has to be treated but the family as well, and until the house is quieted no progress is possible. And it is remarkable that while the early nervous symptoms—sleeplessness, changed temper, headache—are regarded with singular indifference, the first notice of over-movement provokes extravagant apprehension on the part of relatives, the most prevalent fear being, however expressed, that the child will go mad. To quell such alarm needs tact as well as knowledge, but the mere statement is reassuring that not one child in a thousand is permanently injured by the disorder.

Turning then to the patient it is common to find, when chorea is of any severity, that the subject of it is violently restrained with all the force of two or more excited attendants. These having been dismissed, ample bed space should be provided for the fullest movement, without risk of injury. This may be done by placing two bedsteads side by side, or by spreading mattresses on the floor with suitable bedding. The

patient may now fling the limbs about without restraint, and should have full sanction so to do. It is surprising how great is the relief, both to patient and family, that follows these simple measures. Much of the mediæval, besides its name, still lingers in the minds of the vulgar in reference to St. Vitus's Dance, and the effect of dealing with it in a rational spirit is like the breaking of a spell. (See especially Case 13).<sup>1</sup>

Two needs still remain, and in the severer cases we are now considering they are sometimes hard to supply—food and sleep. To feed sufficiently requires address and practice. The main difficulty is to administer timely nourishment in sufficient quantity. For want of perseverance in this duty or from lack of experience, some patients, it is to be feared, are habitually underfed to the aggravation of their malady. It must be remembered that the ability to swallow, like all the other symptoms of the disorder, is subject to frequent variation. The nurse must watch her opportunity. I have never known an instance where skill and patience were finally baffled, but I have seen them sorely tried. In some instances

<sup>1</sup> The distress and demoralisation which chorea commonly excites in its adult subjects are well illustrated in some of its fatal cases (Chap. III.), and in others that are of unusual severity (Cases 12 and 13). There is the widest difference between them and young children who, having no deliberate will and but little experience of muscular conduct, are ready to concur in any kind of movement. These elder patients often believe they are going mad; their friends often ask whether the complaint will 'not go to the brain.' In such circumstances it is no small thing to give the body the amplest opportunity of movement without fear of injury, and to give the mind the best support that can be devised by representing the movements as having a curative design. In extreme chorea this can only be accomplished by the method here recommended—room for movement and no discouragement to moving; abolishing the distress of physical restraint and allaying in great measure the apprehension and even terror which the movements themselves excite. Such violence left unchecked soon spends itself, just as a high sea gradually smoothes on a shelving shore, but rises into greater commotion when some direct obstacle opposes it.



the question may occur of nutrient enemata or suppositories to supplement ordinary feeding.

The procuring of sleep may be not less difficult. Insomnia is an early rather than a late symptom, and it may yield to strict quiet, with close attendance and a darkened room. But if sleep be absent—absolutely absent—over a period of twenty-four to thirty hours, it must be procured by narcotics: chloral or bromide of potassium, or the two together. But every repetition of this needful drugging is to be regretted. It is better to encounter the dangers of sleeplessness, such as they are, rather than to bring about the evil train of symptoms that always follows the prolonged use of any narcotic whatever. At the same time a timely dose such as I have indicated or even hypodermic injection of morphia is sometimes absolutely necessary. Sleep must be had.

In chorea of ordinary severity the needs are less pressing. The patient must in all cases be kept at first in bed, and no open notice taken of movement infirmity. She is to be helped where she needs help, amused but not excited, and attended only by one or two persons. Near relatives are apt to be too anxious and sympathetic, and are therefore less suitable attendants than nurses. The main consideration is to establish a fitting relationship, so to speak, between the child and its disorder, sometimes neglecting the over-movement, sometimes even approving it, and taking special precautions always to set the mind at rest in regard to it.

Not seldom this wholesome rule is disregarded, and the patients are called upon to perform acts beyond their reach—to touch the nose with the forefinger, to clasp the hands, and so on. The mortification of failure in such attempts tends, I am certain, to retard progress.

It is seldom at this early stage that any direction to the patient to control movement is of service. Muscular pro-

priety is never so difficult to preserve as when care is taken about it with the knowledge that it is being watched, and the apprehension that failure will entail some kind of penalty. There is no nicer point in the whole scheme of treatment than to know exactly when to make direct appeal to the child's own volition so as to ensure that its first active exercise of will shall meet with the success which is to strengthen, and not the failure which is to enfeeble it. The observation of the disorder continually teaches how much its progress depends upon trivial circumstances of its own providing, how judicious praise, or some special motive to be quiet, or the attention pleasantly arrested, will procure sometimes a quite sudden amendment; and how by the help and encouragement of this progress a fresh step is made towards recovery.

So soon as this quiet routine of life is well established, when the doctor, as well as the nurse, is regarded as a friend and all outside worry excluded, the patient will be usually on the way to recovery.

It has been said already that the progress of chorea is not accurately described by saying that it gets gradually worse and then as gradually better. Very commonly there is a period—a day or an hour—of which we can say that in it the disorder is got under, not finally vanquished but mastered for the time. And in this uneven course many incidents occur of which we may avail ourselves for the service of the patient. Thus the power of walking, which may be completely lost even with moderate chorea, is often regained very quickly, and the same may be said of writing. Of such improvement the child should have the full benefit in being allowed extra freedom.

There is nothing special to be said of diet. Food should be light and nutritious; and, in view of the muscular fatigue of extra movement, it should be given liberally, and at frequent intervals.

The sufficient and regular action of the bowels is of great importance. The old doctrine that attributed chorea to intestinal irritation and overloading of the lower bowel need not be revived, but there can be no doubt that physicians were guided aright in advocating the use of laxatives. A gentle somewhat free action of the bowels is, in my belief, very important throughout; and in these days there are many excellent drugs suitable for procuring it. Cascara may be specially commended. In the case of young children some material irritation, the occasional presence of undigested food, or of round worms, make it necessary to commence treatment with moderate purgation, and it may be to employ santonin as well.

As time passes and the child improves, limb exercise must be encouraged, and the power of walking carefully tested. I have not found that 'drill' practice is of much service. In fact, amendment once commenced generally proceeds rapidly enough; it is helped on by the child's observation of each day's new accomplishment, and needs little help or stimulation of ours.

Of the incidental ills of chorea there is none more striking than its sudden limb and joint pains, acute, fugitive, apt to be worse at night, the wrists being their favourite seat. Local applications may here be of benefit, but the symptom is so short-lived that there is little to be said of its treatment.

In quite exceptional cases patients at the end of three weeks or a month show no sign of improvement whatever. You must be prepared in such event to have the wisdom of your treatment disputed, and may even have misgivings of your own in respect of it. The parents may become impatient and dissatisfied, urging that something more should be tried, or suggesting 'a second opinion.' So beset, the practitioner is sometimes driven to mischievous activity against his own

better judgment. In the tranquil moral atmosphere of a hospital we are exempt from these worries, and we know that if not subjected to endless experiments these lingering patients do well enough in the end. This tardy progress is found amongst children who are industriously and variously drugged as much as with the others. But in any case, this settling down to a fixed state of infirmity, both muscular and mental, needs a treatment of its own.<sup>1</sup> It may be compared with a similar condition in hysteria, and concurs with the same enfeebled will-power which, by mere force of habit, every day tends to aggravate. Indulgence and allowance are now out of place. The patient must be roused, and mentally stimulated. The kind and the degree of urging will depend upon age, temperament, and intelligence, but provided there be no chronic mental defect, ultimate success is almost certain. (Case 9.)

No other principles than these apply as I believe to that acute form of chorea of which we have examples in Cases 6, 12, and 13. The vital power of such patients begins to fail early, and it fails very rapidly. The violent and ceaseless movements, the utter want of sleep, the inability sometimes to swallow, sometimes to get food fairly into the mouth, it may be even the very measures designed for relief, in active restraint and supervision, concur to produce this result. We are face to face with

<sup>1</sup> Dr. Bastian (*Lancet*, July 13th, 1889) has described the treatment of severe and protracted chorea in a hysterical girl of 20, by *prolonged sleep*. Twenty grains of chloral, with ten of bromide of potassium, were given at each period of waking which exceeded half an hour. Such dose was at first required every four hours, but afterwards every six hours sufficed. The girl was thus kept asleep almost continuously for three weeks. She became extremely emotional after the treatment, and had passing delusions, but in the end did well. The author explains that this treatment by sleep is *applicable only to obstinate cases that have continued for months or years without abatement*, the object being to ensure prolonged cessation of the movements, and so lessen the risk of permanent injury to the motor centres.

a very grave juncture,<sup>1</sup> demanding patience, calmness, and dexterity. In addition to the bed arrangements just spoken of, grateful help may be given in some instances by holding the hands or the arms, or by tightening a sheet over the body (as in Cases 7 and 17), but any continuous or forcible restraint, or coercion of any kind against the expressed wish of the patient, is to be avoided.

The sudden and serious vital depression which these patients often exhibit must be promptly met by precisely the same means as apply when similar symptoms threaten in acute disease. These violent cases do not long remain in one stay. The real remedy to be confidently and continuously relied on is food and all the conditions of rest, mental and bodily. Where there are signs of typhoid sinking, together with insomnia, and other remedies are either not applicable or have been found to fail, then over a short time and for this specific end we are justified in giving alcohol freely.

In such severe cases as these the real pinch of the moral treatment is fully felt. It has more than the disease to overcome. When making use of drugs every fatal case is supposed to occur inevitably and in their despite—every recovering one is due to their agency. But when using such methods as I am describing every fatal case has been left to perish, while every recovery is spontaneous. Moral encouragement and support are common-place remedies not capable of much diversity; but there is literally no end to drugs, and the feebleness of purpose

<sup>1</sup> Dr. Goodhart gives an account (*Lancet*, 1882, vol. i., p. 181) of the treatment of severe chorea in twelve cases by *massage*. It was applied twice daily, for 15 minutes each time for the first seven days, and for 20 minutes afterwards. He observed that in four it had a most marked effect, and was 'influential in saving the child's life.' Three of these four, however, were little boys of an age when it is difficult to show that chorea is ever fatal. The writer commends the massage treatment in acute chorea only. I have myself repeatedly tried it in lingering cases that failed to make progress, but with no striking result.

which appeals to one remedy after another is easily made to appear fertility of resource.

Chorea, as we have seen, will sometimes prove a very tedious and lingering affection, getting better and worse by turns, suffering, it may be, sudden relapse with acute, transient pains in the joints and limbs, and sometimes, in boys as well as girls, violent hysterical outbreaks. The most suitable treatment for such patients is not that which a general hospital is always able to provide. They have there to endure the disturbing and sometimes distressing incidents of ward life, the sight of death or delirium, or of others affected like themselves. On the other hand, I can speak with the confidence of experience, of the service of such unbroken quiet as has just been described. Cases of long-standing chorea sent from the Great Ormond Street Hospital to the Convalescent Home at Highgate, get rapidly and sometimes almost suddenly well. For many years this has been accomplished in almost every instance without the aid of medicine.

Finally, we come face to face with the question of drugs. Apart from narcotics (whose use is limited to severe cases), and apart from ordinary medicaments, such as aperients, is there any drug directly serviceable in the sense of having some specific action? In my belief there is none. True there is no disorder than can more easily be made to appear as if influenced by drugs than chorea. It has a natural tendency to recover, and once on the right road, recovers rapidly, while the patient is often largely influenced by any confident prediction of amendment. Provided it be given at the right juncture with the requisite assurance of relief, there is little left for the drug to do. Many esteemed remedies for chorea are now no more: belladonna, sulphate of zinc, antimony, conium, and others. Arsenic has deposed them all, and will no doubt itself in its turn lose popularity and give place to something else.

But doubting the specific virtue of arsenic in this complaint is not the same as to deny its influence. I have said that good health is not inconsistent with chorea. But it cannot be denied that bad health, the failure of health, is sometimes the signal for its cessation. Acute illness of many kinds will have this effect. It is very striking, for example, when a choreic patient contracts enteric fever, to witness the movements of the one affection exchanged for the characteristic stillness of the other. It is reasonable to suppose that many poisons besides that of enteric fever act sometimes in a similar way. In other words, if arsenic be 'pushed' (as we euphemistically say), that is, if it be employed in poisonous doses, we might expect by analogy that the choreic subject would sometimes be reduced to stillness by being made ill. But it is a large price to pay for a result which even so is only secured now and then. It by no means follows that so soon as the well-known symptoms of arsenical poisoning appear chorea will disappear.

Yet there is more to be said about drugs than this. It is impossible to ignore the fact that many competent observers have given testimony in no doubtful terms in favour of drug remedies for chorea, especially arsenic. A constant habit of drug giving may no doubt conceal from some the almost equally constant habit of some affections to get well without their aid; but when all sources of error are excluded, imperfect observation or mistaking recovery for cure—how is it that the very same preparation and dose should be uniformly curative in one set of cases, and uniformly inert in another? Consult the affection itself for the answer. Faith makes the great, the real virtue of these medicines. It is but a modern illustration of what was more grossly shown in the early history of chorea. Its subjects, now as then, reflect at once the temper of those around

them, and drugs confidently given are of real efficacy in consequence.<sup>1</sup>

The treatment of convalescence, it has been said already, is generally a lengthy process. For a considerable time after apparent recovery much care is necessary lest the child should be exposed to any influence similar to that which occasioned the first attack. Too early return to school, the resumption, when still physically weak, of manual labour, even a sudden recalling of all the circumstances of the seizure by being brought back to the place where it happened may revive a chorea which has disappeared for weeks.

In conclusion, if I were to express in few words my own opinion as to the treatment of chorea—the treatment I have myself adopted for many years in hundreds of cases (and this is my warrant if I speak dogmatically)—it would be to say that it consists in sufferance, patience, cheerful anticipation, a firm unshaken purpose, self-reliance—the very same virtues we seek to arouse in the patient. With the vast majority of cases there is no ground whatever for anxiety, and whether the children return to perfect propriety of movement a little sooner or a little later, what does it matter? The more your disquiet the longer the child's disorder. As for the exceptional and severe cases—though we have examples to warn us that

<sup>1</sup> Those who suppose children slow to reflect impressions produced upon their elders by causes which the childish mind cannot be supposed to comprehend may profitably read Wesley's 'Journal' as to the effect of the revival preaching of his day. 'I heard,' reports an eye witness, 'many cry out, especially children, whose agonies were amazing. One of the eldest, a girl of 10 or 12, was in violent contortions of body. Among other children who felt the arrows of the Almighty, I saw a sturdy boy about 8, who seemed in his agony to struggle with the strength of a grown man. I observed besides one little girl deeply convinced, and a boy nine or ten years old,' &c., &c. See Lecky's 'England in the Eighteenth Century,' vol. ii., p. 583, *seq.*



they are not absolutely free from danger, yet these examples (or some of them) teach that this danger, be it more or less, is partly from the chorea and partly from the sometimes reckless means employed to suppress it. Violent remedies—remedies themselves dangerous to life—are ill suited to a disease that of itself, however violent, habitually recovers.

It may seem too sanguine to expect any general acceptance of such views. A natural instinct of mankind craves for remedies. It is improbable, therefore, that chorea will ever be formally exempted from the number of disorders that drugs can cure. Nevertheless, considering its nature, sources, and subjects, I believe that the treatment indicated throughout this work is in principle right, and I make bold to add, as a matter of fact, that in practice it is not unsuccessful.

## CHAPTER XI.

## ILLUSTRATIVE CASES. COMMENTS.

THE following abstracts, classed in order of age, are intended to exhibit some of the characteristics of chorea, *e.g.*, its connection with rheumatism and with endocarditis, dependence on nervous causes, variations with the time of life, cardiac and paretic symptoms, limbs chiefly attacked, occasional chronicity, occurrence in pregnancy, together with methods of treatment. The cases are referred to in the foregoing pages in connection with the points they are supposed to illustrate, and are given with more or less detail and comment according to the circumstances. The selection having been made with a view to exhibiting certain special symptoms cannot be taken as a general picture of the disorder, such as would be afforded by a like number of cases taken consecutively. It will be noted especially how large a proportion are beyond the age at which chorea is common; and how, with these elder subjects, the emotional element becomes prominent. With the exception of a case quoted from Dr. Brabazon (Case 12) as a more striking example of the supposed occurrence of chorea from imitation than I could myself quote, all the patients were under my own observation.

CASE I.—*Heart Disturbance arising in a child while under observation in hospital without distinct evidence of Rheumatism, and shortly followed by General Choreia.*

Alfred G., aged 10, was admitted into the Children's Hospital July 18, 1880, with an account of six months' loss of flesh, and cough, pain

in left side and loss of appetite for the last fourteen days. Neither his parents nor he had had rheumatism. The heart sounds were natural. Three days after admission he had some tenderness and pain over the right sterno-clavicular articulation. Temperature, however, had not risen over 100°. There was now audible over upper sternum a well-marked (basie) *diastolic* murmur. In the course of the next week his temperature became normal, and no fresh symptoms appeared. But now the heart's action was irregular and 'tumbling,' the murmur persisting. The boy, appearing well as to his general health, was sent to the Convalescent Hospital at Highgate. There the action of the heart became thumping, and much accelerated. With this, a restless movement came on which soon developed into a general chorea. His parents, seeing him at this time, and alarmed at his appearance, insisted on taking him away, and he was thus unfortunately lost sight of.

CASE II.—*Heart Disturbance following fright, with pain not identified as rheumatic, and very slight and transient Chorea.*

Emma D., aged 10, was admitted May 29, 1878. A month before she had been much alarmed by being bitten in the hand by a dog. Two weeks after she complained of pain in her knees. On admission there was still pain, but neither redness, tenderness, nor swelling of joints. For three days her night temperature was raised, and there was slight choreic movement of arms and hands. A soft systolic apex bruit was audible. As the pyrexia disappeared this soft bruit developed into a loud blowing mitral murmur.

[These two cases are samples of many where the heart symptoms preceding the development of chorea are connected with local pains and slight pyrexia, both of short duration, and making probable the belief that both cardiac murmur and chorea are of rheumatic origin.]

CASE III.—*A Moderate and Partial Chorea of LEFT Side following rheumatism converted, upon the occurrence of fright (more than three months after the rheumatism), into Violent General Chorea, in which the RIGHT Side was most affected. Succeeding joint affection.*

Louisa P., aged 10, a delicate-looking lean child, had had rheumatism five months before admission, followed (after a clear interval of three weeks) by twitching movements of the left arm and leg. This disorder

continued up to a week before admission (*i.e.*, for about three months), the child getting about as usual. She was at this time greatly alarmed by being beaten by some boys in the street. The movements thereupon became general and violent, and worst on the *right* side. Chorea was still severe on admission, and for the first three weeks she had to keep her bed. She made good gradual recovery, and was fairly steady in about five weeks.

When thus on the way to recovery this child happened to fall on her right elbow. The joint next day became swollen, red, and tender, and was put up in splint in flexed position. The swelling and redness shortly subsided; the elbow joint, however, remaining stiff and the arm constantly maintaining a position of rigid flexure, the patient was referred to Mr. Macnamara. It was found that under chloroform the joint movements were normal, as they appeared to be also in sleep, and Mr. Macnamara was of opinion that the elbow joint was perfectly healthy. The rigidity, nevertheless, persisted. Considerable force, met by continuous and unvarying resistance, served to flex the fore arm partially; the girl giving utterance to pain the while, but without any hysterical manifestation. By the time these symptoms had developed, the chorea had disappeared, and the child's general health was good. Not only could the arm be easily flexed during sleep, but the condition improved so rapidly upon the promise to the child that she should go home when the joint was well that, although the extreme resistance to flexure and other signs were anomalous, the symptoms generally coincided best with so-called hysteric joint.

[The case is introduced to show how a slight localised movement may be aggravated to severe and general chorea by nervous shock. It shows also, like Case 10, that chorea in its second attack may change its side. Further, the joint affection which arose after a fall in the course of convalescence is a good illustration of what may be called the joint sensitiveness of chorea and of the sort of symptoms which pass as rheumatic upon very dubious grounds.]

CASE IV.—*Chronic Chorea in a boy of eleven, ultimately fatal.*  
*Mitral Endocarditis.*

[The following case was under the care of the late Dr. Fuller at St. George's Hospital, in 1864, and reported by myself, then Medical Registrar, the *post-mortem* examination being performed by Dr. Dickinson, then Curator of the Museum.]

Leopold L., aged 11, admitted July 13, 1864. The boy had before been an in-patient, and was discharged improved, but on returning

home his disorder at once returned. At the time of his second admission it had lasted three months. No cause could be traced. The patient was well nourished, of dark complexion, and not unhealthy aspect. The chorea was general, but at the first not extreme. He was given iron and sulphate of zinc, and for a short time improved. This early amendment, however, not continuing, he was ordered strychnia with iron, and morning shower baths. At the end of five weeks from his admission his state had become distressing. He got little sleep, had wasted much, the tongue was frequently bitten, and the skin scratched with the finger nails.

The liquor arsenicalis was now given in five minim doses, thrice daily; many other drugs followed, particulars of which it is unnecessary to insert, since none exercised the slightest beneficial effect. The symptoms progressed without interruption, the skin was excoriated, the tongue ulcerated, and bed-sores began to threaten. At this advanced period of the disease it is noted, 'There seems a strange mixture of temper with the disease, rendering it difficult to discriminate between the two. His most violent paroxysms may be controlled for awhile by speaking sharply to him.' Ten days before his death he was tied down, and, to prevent further injury from his scratching, the hands were secured. So he lay like one possessed, struggling and kicking, the mouth and tongue deeply ulcerated, and the body lacerated and bleeding wherever his fingers could reach it. He died September 17, after over two months' residence, and more than five months' duration of chorea.

*Post-Mortem Examination.*<sup>1</sup>

The body was much emaciated, and the skin rubbed off the prominences of the back; the hair was worn off the back of the scalp, and under its skin was a collection of pus; the lips were scored with fissures. The *brain*, both grey and white, was injected, the ventricles natural.

The *cord* showed on its surface large vessels distended with blood. On section in various situations, its appearance was not unnatural; but on closer inspection portions of the grey matter seemed duller and more yellow than natural, particularly towards the upper part. External to the membrane were much distended veins lining the vertebral canal. No microscopic examination was made.

The *heart* at the inner edge of the mitral valve showed a line of soft beads of lymph easily detached. The left ventricle was contracted.

<sup>1</sup> This case is referred to in Dr. Dickinson's paper, *Med. Clin. Trans.*, vol. lix., and included among the 22 fatal cases he has collected.

The kidneys were congested. Other organs natural.

[Although incomplete in some respects, I am induced to select this case for quotation in this place, owing to its extreme rarity. Chorea in a chronic form occurring to a boy of this age, and persisting up to the time of death, *no extra symptoms supervening, but the disease maintaining its proper characteristics to the very end*, is, as Chapter III. shows, an excessively rare event. There was no trace of paralysis, no known rheumatic connection, and no cerebral symptoms beyond those of temper. See Chap. III., p. 57.]

CASE V.—*Temporary Heart Disturbance (arhythmia, tumultuous action, mitral murmur becoming presystolic, concurring with severe Chorea in a child recovering from Rheumatism, the immediate cause being fright. Advantage of free movement.*

Ellen B., aged 11, a bright, intelligent child, in the sixth standard, well nourished, but very 'nervous,' was admitted (Dec. 3, 1892) with severe chorea due to a fright, caused by a mouse suddenly appearing from the wainscot when the patient was recovering from rheumatism.

The movements which were considerable on admission became violent the following day and the child slept but little for the first three nights. Speech was jerky and difficult. There was at first a blowing apex murmur, but the heart was quiet and regular.

A marked benefit resulted (as in Cases 7 and 17) from placing cots together so as to afford ample room for free movement. Drugs were necessary to procure sleep, and chloral was more successful to that end than the bromides.

On the second and third days of her admission, when the severity of the disorder was beginning to lessen, the heart became markedly irregular and tumultuous. On the fourth day it quieted in these respects, but a distinct presystolic murmur was now added, thrill doubtful.

After the first four days the chorea rapidly decreased, and at the end of a week the child was replaced in a single bed. As regards the heart, the presystolic murmur became fainter and presently inaudible. On discharge after two months' residence, well, the heart was quiet, the apex murmur heard at first was now indistinct, and there was no presystolic sound.

[This case, besides illustrating the good result following freedom of movement, exhibits heart symptoms often accompanying chorea, both in

rheumatic and non-rheumatic subjects, and due presumably in both cases to an endocarditis which is recovered from. In the present instance the child's history makes it probable that the endocarditis was rheumatic, and the ultimate prognosis as regards the heart is less favourable on that account.]

CASE VI.—*An Example of Chorea, exhibiting rapid transitions and dependence on mental Condition.*

Elizabeth D., aged 12, was admitted on the first occasion for chorea brought on by fright on being threatened with corporal chastisement; the movements, which were not severe, commenced in the left leg and extended to the arm and hand. After a short residence in hospital she was discharged nearly well, but returned after three weeks worse than at first. This second attack had followed upon a blow. The child walking carelessly had struck her head violently against a lamp post. The movements now concerned the hands, head, and face; there was likewise much restlessness of the tongue and difficulty in speech. She improved considerably after four days in bed, was then allowed to be up for part of day, and could move about with ease and comfort.

Two days later (a week after admission) and a few hours after she had been much disturbed by the news of the serious illness of her father, she complained of pain in the right arm, wept, refused food, and occasionally vomited. All the movements were now much increased; the face and *right* side being the most agitated, and speech very difficult. Night temperature was 100°, the pulse not frequent. Her position, lying low down in the bed, with restless working of the mouth and considerable limb movement indicated at a glance a severe degree of chorea. In attempting to clasp her hands she would make movements quite distant from her purpose, but once or twice, as by a sudden jerk, succeeded in bringing the palms accurately together. The same sort of intermittent co-ordination was observable in other movements. She was unable to maintain a grasp, and much arm-agitation was occasioned by attempts to take food. Here, indeed, was the main difficulty. It was only after much patience and many trials that food could be swallowed, attempts at deglutition being often followed by general agitation. She was acutely sensitive, readily shed tears when noticed by strangers, and had the usual emotional exhibitions of crying and laughing common in hysteria. These symptoms continued for three days, at the end of which time, owing probably to the inadequate amount of food taken no less than to muscular fatigue, she was much weakened, and ominous signs of sinking began to threaten. Brandy was now given. It was by

sudden jerky effort, similar to that which impressed all her movements, that she succeeded in swallowing.

Upon this she slept more soundly than she had done for some time ; and was still sleeping when suddenly awakened by a street band. Thereupon the muscular agitation returned with almost maniacal vigour, the girl screaming and crying, but unable to utter words. This fit spent itself in about an hour, and the patient, much exhausted, again slept. From this the improvement was rapid and continuous, and the difficulty in swallowing almost disappeared, yet she was still quite unable to walk. The walking power was almost suddenly regained ; on February 10, a month from time of admission, she was convalescent.

[This case speaks for itself, in its rapid transitions, emotional admixture, variable power of co-ordination, early appearance of alarming prostration, sudden relapse, difficult management and response to treatment. In some of these respects it may be compared with the one to follow.]

I need only append the following note by the Sister of Harpur Ward, who most carefully and judiciously directed the nursing of the child :— ‘All through her illness the girl has been excessively sensitive. We had to be most careful how we talked in her hearing. A misunderstood word would bring on difficulty of speech.’]

CASE VII.—*Severe Chorea with Muscular Paresis ; variable ataxy ; extreme agitation of the hand ; benefit to be derived from mechanical support and muscular exercise.*

Annie M., aged 12, an intelligent and tractable child, lean but of healthy aspect, was brought to hospital with rather severe chorea of about three weeks' duration. The affection was first noticed from her dropping her glass while drinking. The over-movement was now general, but affected the right side chiefly. Both hands were very unsteady, and the right could not be fairly extended from the wrist, but habitually drooped more or less. She was quite unable to stand or walk, or move the legs in any desired direction. The girl had never had rheumatism, and the attack (which was the first) was attributed to falling downstairs, or, perhaps, to toothache and extraction. There was neither chorea nor rheumatism in the family history. The heart was slightly uneven in action, and the first sound prolonged, but without definite murmur.

The choreic movement soon became much worse, so as to require special watching. With the full consent and concurrence of the child



means were employed mechanically to restrain the limbs. A sheet wide enough to cover the whole trunk was stretched over her and tied round the bed. This was applied and removed at her pleasure, and by its aid she got sleep and soon improved in steadiness. But now with the lessening of the more active agitation a general muscular weakness became apparent. The girl lay low in bed, and the inability to stand was now due rather to want of strength than want of steadiness. The rhythm of the heart had become irregular. There was no appreciable wasting. It now became a nice question whether her condition might be best met by further rest in bed or by directing such use and exercise of the muscles as was possible. In the belief that the paresis was parallel to that met with in hysteria, and with a view to testing that opinion, the girl was directed to be up for a few hours daily. In the course of a fortnight she was able to walk with some support, dragging the legs somewhat, but showing no mis-direction. The face was still unsteady, and, as at the first, words were uttered with difficulty and in jerks; but the stress of the disorder was now chiefly in the right hand, which was utterly choreic, and unable either to be still or to make any attempt at grasping.

In pursuance of the same plan that had succeeded with the general disorder, this hand was now bandaged down on a straight splint, the child concurring in this as readily and, indeed, eagerly as in the case of the sheet. For the first week of this treatment the hand could be felt with the fingers, wriggling about under their bandage. After this the hand became rapidly steadier. She was able first to hold a pencil, and soon to write, *the return of writing power being noted to a day, and even to an hour, while once regained (as in the case of walking), it was very soon regained completely.* The girl was in hospital altogether about three months, the first fortnight of that time showing a severe degree of chorea, and the rest of it being occupied with recovery, which, although slow on the whole, was rapid and almost sudden in its main incidents.

[Besides other points for notice already alluded to in the body of this work, this case may be quoted with some others to show the benefit of restraint. The mechanical aids to steadiness which were given to this girl, if they did not quicken recovery (and I am not certain that they did), were yet grateful to the patient, and were applied only in so far as they were so. Restraint which is not directed and qualified in this way, restraint sternly ordered by the doctor without consulting the patient, is more likely to do harm than good, and sometimes does very serious harm. But with the patient's concurrence, and with a clear

understanding that it may be used and disused at will, mechanical restraint may sometimes be made use of, on and off, with manifest advantage, even in cases more violent than the present. Another point of interest concerns the treatment of the general paresis, and the need of stimulating the muscles to exertion. The rapid transitions of the disorder, and especially the quick recovery of the use of the fingers shown in the present case, are highly characteristic features.]

CASE VIII.—*Chorea following Acute Rheumatism and Endocarditis, and without obvious nervous cause.*

George H., 13, a shop boy, pale, slightly built, and tall for his age, was admitted into Westminster Hospital, October 20, 1880, with marked chorea of hands, mouth, eyes, and tongue, the head and legs being less affected. There was but little muscular inco-ordination. Pulse was 90, temperature normal. The heart's impulse was visible over a large area in normal position; there was a thrill perceptible of varying distinctness, and a blowing systolic apex murmur which was conducted to axilla and left vertebral groove. Pulmonary second sound accentuated.

This boy was stated to have been strong and well till about a month before admission, when he had acute rheumatism affecting most of the joints. He was laid up for twelve days and attended by Mr. Lattey of Kensington; he had præcordial pain together with the physical signs of endocarditis. Recovering as was believed from this attack, he went back to his business at Richmond. Three days later he was again attacked with joint pain, this time attended with choreic movement. There had been no previous rheumatism and none of his family had had rheumatism. No mental cause of any kind could be made out. The boy throughout was perfectly composed. There was nothing of significance in his family history except that his paternal grandfather had died insane.

The boy was kept in bed, but no active medicinal treatment was adopted. In less than a week he had recovered fair use of legs. His full recovery was somewhat retarded by a painful whitlow, but within three weeks of his first admission he was almost steady. He was remarkably cheerful and unemotional.

In this case the rheumatism and the endocarditis are unquestioned, and there is no room for supposing any nervous upset. It shows therefore, some real connection between rheumatism and chorea. The patient was probably sent back to his ordinary work too early in his convalescence from rheumatism. (See Case 11.)

CASE IX.—*Chronic Chorea with incontinence of urine, emotional attacks and temporary Heart Disturbance. Condition of fatuity succeeding. Improvement with discipline.*

Samuel S., aged 13, a fresh-coloured healthy schoolboy, of blunt intelligence, had pains in the left thigh ten months before admission, shortly followed by choreic movement of the same side. The present is a second attack of three weeks' duration, affecting the left arm and, especially, the face (the forehead and eyebrows being in constant movement). There is no apparent cause for this second attack. The case, never violent, was extremely obstinate. It was marked by three notable incidents: incontinence of urine; emotional or hysterical attacks; and great variability in the character of the heart's action. The incontinence continued more or less throughout his stay. The emotion was exhibited on the occasion of a sudden death in the ward, when the boy had a fit of crying and sobbing with much increase in the choreic movement. The heart, at first regular and without obvious murmur, became for a while irregular or uneven, and developed a variable murmur which exercise made louder; it was post diastolic in time, and heard best just above and within the apex impulse.

After many variations this boy fell into a half fatuous state, but, as will be seen from the description, it was extremely difficult to estimate the precise mental condition. The choreic movements, moderate but general, were greatly aggravated on moving, and he was quite unable to walk or stand. He lay habitually on the back, low in the bed, constantly grimacing, and passing urine involuntarily. When addressed he would sometimes merely grin, and sometimes make an effort at speech, never getting out more than a word. He took food well, but wasted somewhat. His temperature was habitually sub-normal, between 97° and 98°, pulse very variable, ranging from 96 to below 70.

When this boy had been over three months in hospital it occurred to Dr. Dorkin (who then had charge of the ward and to whom is due the full credit of this treatment) to make trial of moral persuasion to exertion more forcible than had hitherto been employed. The patient was with some difficulty got out of bed, and unavailing attempts were made to get him to stand with the help of some support. This failing, a faradic current of some strength was applied, much to the patient's dismay. *The next day he stood*, and from that time made continuous and rapid progress, no further severity of urging being called for. The face unsteadiness in lessened degree persisted, but in other respects the

boy was almost well three weeks from the commencement of the active treatment.

[The lesson of this case, so far as treatment is concerned, speaks for itself. Yet, though it yielded rapidly to moral suasion applied at the right time, it was truly a case of chorea as evidenced by the continual movement of the face no less than of the limbs, by the speech difficulty and the condition of the heart. The variability of the pulse and sub-normal temperature were further signs which could not be mistaken. What the treatment accomplished was to break the monotony at the time when the affection, no longer active, had left a stupid boy without the vigour to bestir himself unless strongly roused. Such treatment is only to be commended in similar cases, yet its success here is one of many illustrations of the points of contact between chorea and hysteria. It need hardly be added that faradism has no specific effect, and that any other application equally disagreeable would probably be as effectual.]

CASE X.—*Choreic Movement of the Hand following Partial Paralysis of the Arm after heavy lifting. Choreia changing side.*

George C., aged 14, a healthy boy, kitchen-help at an hotel, admitted April 15, 1881, stated that four weeks before he had lost power in the right arm after carrying heavy trays. A week after this loss of power, and first brought to his notice from his letting a cup fall, the boy got choeric movement in the right arm and hand. He had had no rheumatism or previous illness. Heart was natural. During his residence this boy got a passing attack of pyrexia, when he exhibited general restlessness, which for a few days threatened to pass into severe chorea. It was short-lived, however, and on its subsidence left him unsteady in the face as well as in the affected arm. He was made out-patient when still imperfectly recovered.

This youth was admitted a second time, with chorea which was now most on *left* side.

[This case shows the association of chorea with that functional paralysis which arises from fatigue of certain muscles, the subject, as the subsequent history showed, having a predisposition to chorea. The seat of the chorea on the second admission affords one of the many exceptions to the rule that subsequent attacks of the disorder are repetitions of the first.]

CASE XI.—*Chorea from overwork after Convalescence from Rheumatism with emotional admixture, delusions, &c. Relapse, with recurrence of Rheumatism. Muscular Paresis occurring together with mitral murmur.*

William L., a healthy-looking youth, aged 16, a railway porter, admitted April 7, 1881. After an attack described as acute rheumatism (and for which he kept bed two days, and remained at home by the fire for fourteen days longer), this youth, on convalescing, was put to new and harder employment as a railway porter. He kept at this, although feeling overdone, until a fortnight before his admission, the joint pains not wholly leaving him. He then, while still at work, had an hysteric fit, crying and kicking, on recovery from which both hands were found to be choreic. Next day the legs were affected, then the head, and in a week's time the speech was difficult. The pains increased as the chorea appeared.

On admission there was choreic movement of both hands, and some dragging of the left leg. The hands worked awkwardly, and their grasp was insecure, the right hand being the worst. There was much speech difficulty, and the face was vacant, partly no doubt from muscular fault, although besides the youth was wanting in intelligence. The heart sounds were at this time normal.

After about three weeks' residence, getting about and feeding himself with difficulty, this boy had a severe attack of general chorea with frequent shrieking and temporary delusions; as that his brother had met with an accident on the railway. With this attack the wrists became painful and tender, and, on the partial subsidence of the pain, the right wrist dropped, insomuch that for a while the hand could hardly be raised into line with the arm, the grasp was likewise very feeble. Along with this a systolic murmur appeared most audible at the base in the pulmonary area. In two days and quite suddenly the wrist power returned, and the cardiac murmur disappeared.

The boy remained in hospital for some time longer, able to get about, and in good general health, but still unsteady, especially when the hands were employed. He was ultimately made an out-patient.

[This case illustrates the occasional alternation of joint pains (in this instance probably rheumatic) with chorea; also the occurrence of a distinct paresis of certain muscles along with an equally distinct cardiac murmur, both symptoms being temporary. The exciting cause of chorea in this instance was a not uncommon one, the overstrain of harder work than the youth was accustomed to when imperfectly

recovered from rheumatism. The hysteric fits and mental aberration are noteworthy in connection with the age.]

CASE XII.—*The Chorea of Imitation and Sympathy.*

The following case is related by Dr. Brabazon, physician to the Bath Mineral Water Hospital, in a paper upon the treatment of chorea with the Bath mineral waters, published in the *British Medical Journal*, May 15, 1880:—

‘The patient, a girl aged 18, was admitted to hospital for chronic rheumatism and amenorrhœa. She was a stout healthy-looking girl. Her home was in the quiet village of Wrington, in Somersetshire, her mode of life and general surroundings were most unsensational. There happened to be in the same ward with this girl a child suffering from chorea, but not of an exaggerated type. After being in hospital about a month this girl was suddenly seized with chorea in its most severe form. In attempting to walk, she knocked herself black and blue against furniture or whatever came in her way; she could not hold a cup in her hands, or feed herself, and, at night, was sometimes jerked as it were out of bed. Her facial contortions and grimaces were horrible to behold. No votary of St. John, no disciple of St. Vitus, no victim of tarantism could have proved a more perfect specimen of ‘Chorea Sancti Viti’ of the middle ages. It became necessary to isolate this girl completely from her fellows, some of whom, in accordance with the history of the disease, would in all probability have been similarly affected, and the Mineral Water Hospital would have been assimilated to an asylum for ‘jumpers’ or ‘shakers.’ This patient was placed in a room by herself, with one attendant, the room being so padded and arranged that she could not injure herself. Mr. Coppinger, the resident physician, has favoured me with notes of the case. I should mention that in this, as in all the cases which I have seen, the thoracic movements were bilateral. The medical treatment employed was bromide of potassium and chloral hydrate in full doses at night. When the symptoms were most violent, six leeches were applied to the inside of the thighs; and after a time, when the acute symptoms had subsided, she had tincture of belladonna, perchloride of iron, and a reclining bath daily at 100° Fahr. This was palpably a case of acute chorea, induced by morbid sympathy, in a female at the age of greatest nervous susceptibility, and under circumstances of functional derangement adapted to aggravate the symptoms. I think there could be no better example than this of a case of acute chorea, and no case better suited to form a link in the chain of connection between the epidemics of the middle ages and the sporadic cases of the present. It

also bears out to some extent the theory of Dr. Sturges as regards this and other analogous diseases, regarded by him as simply an affection of function.'

CASE XIII.—*Violent Chorea in an Adult, with delusions, followed by Hysteria. Rapid recovery.*

Marian R., aged 20, a fair-haired, healthy girl, living at home in training for a governess, was admitted into the Westminster Hospital with severe chorea. She had had three previous attacks—the first, occurring at 13 when studying with the view to teaching, was due immediately to fright caused by a boy throwing a dead kitten in her face. Thereupon she began to twitch the facial muscles, and became 'excitable' and 'nervous.' Five or six months before this she had complained of what were called 'growing pains,' wherein her legs and joints were affected, but not red or swollen. The second and third attacks occurred near together two years after the first, and when she was 15. They were both of mild character, and little could be remembered of them. The girl had been healthy in childhood, but apt to walk in her sleep. The family history was of a negative kind, and especially wanting in any particulars of rheumatism or nervous disease.

Three months before admission the patient was in bed for a week for supposed rheumatism. Some fortnight later, when still feeling the effects of this illness, she was much alarmed by having to spend a night in the same house with a woman who was subject to fits of insanity, and who associated the patient with her delusions. From that time up to her admission the girl felt 'as if going mad.' She had visions of rats and mice, was always worse towards night, and feared to sleep alone. She would occasionally ramble in her talk, and foretold that she would presently have a return of her old disorder. The actual attack commenced thirteen days before admission, and had been getting worse daily.

On admission the patient was found to be the subject of violent chorea, affecting chiefly the face, speech, arms and hands. A loud systolic bruit was audible at the heart's apex. On being put to bed the movements became much aggravated, and towards evening were so energetic that she was kept in bed with difficulty. She was given bromide of potassium with chloral during the night, but got barely two hours of disturbed sleep. On the following day (June 23), at the time of the usual visit, the jactitations were extreme, so as to require constant watching and holding. She spoke indistinctly, and was fed with difficulty. The pulse was now 108 and intermittent, tongue natural.

Hearing and sensitiveness were acute. As evidence of this it may be mentioned that she was especially disturbed by overhearing a whisper to the effect that her father was not to see her. At times she had delusions in reference to those about her, but these had always a basis of reality, and she was easily recalled to herself. She reached her worst on the night of this second day, and remained in the same critical condition till the afternoon of the third day (the 24th), when it was thought necessary to place her in an isolated ward.

From the evening of the 22nd to the morning of the 25th, this girl required the active service of one or more nurses to keep her in bed and prevent her doing herself injury. The jactitation of arms and body was more than of the legs. There was no emotional excitement at this time, but much anxiety and apprehension. When almost at the worst, and desiring to see her mother, she was able so far to control her hand for a moment as to write a few legible words. It was observed also that whenever the violence of movement somewhat relaxed, she would snatch the opportunity to employ her hands to some purpose of the moment. She could also offer her hands to be held and was partially steadied for a while by such means. Throughout, so far as observation was made, she retained such imperfect power of control as enabled her to moderate her movements for a moment when so bidden, but their violence was heightened by such efforts. The pulse was now 120, the temperature could not be taken at the full strength of the paroxysms, but immediately before (on the 23rd) it was 99°.

So soon as the case had actually assumed the degree of violence that it threatened from the first, it was resolved to adopt no specific drug treatment, but to direct attention to removing all removable discomfort; to supplying food; and to the procuring during night time of a sufficient amount of sleep by the help, if necessary, of narcotics. With this threefold object several mattresses, bounded by pillows and bolsters, were placed on the floor of the room to which she had been removed in such manner as to form a capacious bed ample enough for the patient to lie at length in any direction and to fling her limbs about without injury. She had already had repeated doses of morphia subcutaneously without effect, and on the morning of the third day (the 24th), some hours before her removal to the separate ward, had been given thirty grains of chloral. She got, however, only short snatches of sleep, was fed with extreme difficulty owing, not to any impairment of deglutition, but to the violence of movement, and her strength was beginning to fail.

It was now decided to suspend all medicine whatever during the day, but that, if absolutely sleepless during the night, a hypodermic injection



of a quarter of a grain of morphia should be given. Three ounces of brandy were ordered in addition to such liquid food as she could be got to swallow. The girl was left almost free and unrestrained as regards movement, with the assurance that she might throw herself about without fear of injury and that such agitation was not in itself evil but rather to be suffered as working out the cure. This same night, in pursuance of the plan described, she was injected with a quarter of a grain of morphia, but was restless and sleepless, although less violent. On the 25th the agitation had moderated considerably but increased towards evening. That night the sleeping draught consisted of thirty grains of chloral, a dose which (without absolute need) was repeated on the night of the 26th.

For in fact the morning of this, the fifth day, saw the end of the chorea in its violent and threatening form. The patient had slept for six hours. She now for the first time exhibited emotional excitement in tears and lamentations, the hands and arms, heretofore violently agitated and still unruly, being now employed to some extent in giving expression to these feelings. This hysterical state lasted only for the day. The girl was brought back to the general ward with but slight chorea affecting chiefly the hands. It was now for the first time possible to judge of the patient's aspect and bearing. She appeared to be an intelligent girl, cheerful and contented. During convalescence *she had for two days pain with some tenderness in the left wrist and back of the hand.* On the 27th, the 6th day from admission, she was up and about, some choreic restlessness being still perceptible, but to such slight degree as to make it unnecessary to keep her much longer an in-patient. The cardiac murmur still continued, although much softer and less distinct than at first.

[The points for consideration in the case concern its distinctive characters; method of progress and recovery; and response to treatment.

Here was obviously a chorea due to serious alarm in a predisposed person with the interval between exciting cause and actual outbreak of motor disorder occupied by mental disquiet. It was in many respects different from the chorea of childhood; more severe, mixed with mental disturbance, and for a while threatening to end fatally. The violence of muscular movement could hardly have been exceeded. Yet it exhibited an element of voluntariness. The girl could remain comparatively still for a second or two when so bidden, an increased violence of movement following such effort.

It is to be observed how, in its utmost severity, the disorder of chorea keeps itself clear of spasm. At no time could it be said in the

present instance that any particular limb or set of muscles was in true convulsion. Looking at these patients on the whole, as common observers do, the amount of their disorder seems hardly to be surpassed, but looking at one limb only or one set of muscles, its movement is seen to be neither convulsive, nor repeating, nor even continuous. The arm, for instance, is flung about now in this direction and now in that, with a half-recklessness which is always under some sort of command, and which is considerably modified for the short time that attention can be centred on that particular part. Anyone who will hold the hands of a violent choreic patient for a short time will recognise this diversity of movement. This same absence of spasm is seen further in the grimacing of the face, while the measure of volition is shown in the rarity with which the tongue gets severely bitten or the fæces discharged, as happens so commonly in epilepsy.

While the course of the chorea in the present instance was marked throughout, as it so often is with adults, by great sensitiveness and anxiety (p. 55), it was only when the muscular disorder began to decline that the hysterical symptoms appeared. We see precisely the same sequence in hysteria itself. During the height of hysterical spasm emotion is in abeyance, but when the convulsion ceases the patient breaks into tears or laughter. Thus, notwithstanding the very real distinction between these disorders—the distinction, namely, that the choreic woman is dismayed and distressed at what has come upon her, while the hysteric surrenders herself willingly to a spasm which is in itself a relief—there is an obvious alliance between the two. The older choreic patients are mostly hysterical. The chorea of childhood often becomes the hysteria of womanhood. It may even be difficult to decide in some instances whether to call a restless movement by one name or the other.

A word as to drug treatment. Drugs were pretty freely used at the first for sleeplessness—morphia and chloral—and were of some efficacy, but the real improvement set in when they were stopped and when bodily freedom and moral support were substituted].

CASE XIV.—*Left Hemichorea implicating lower part of left face, hand and foot, in a young pregnant woman newly married, a month after rheumatic attack.*

Jane C., aged 21, ironer, four months married, and in third month of pregnancy. Two and a half months before admission she had had pains, swelling and tenderness in the small joints of both hands and the left foot, for which she kept bed a week. Five weeks before admission (*i.e.* something over a month from the supposed rheumatism and when in

good health) suddenly, and from no apparent cause, she felt a twitching or turning outwards of the left hand, the left foot soon participated, and the left corner of the mouth had a downward twitch. These movements were very perceptible on admission, although not violent. The woman had never before had chorea, and, except for headache and neuralgia, knew of no previous illness. There was no rheumatism or neurosis in her family history. A variable systolic bruit was audible between apex and base of the heart, sometimes loud, sometimes quite absent. The patient improved in a few days, and left the hospital after a fortnight's residence, nearly steady.

[Here was a strictly one-sided affection, involving both face and hand, and foot, in just such manner as irritation of a motor centre would, and without the slightest participation of the right side. Yet it was not a continuous restless movement, but an occasional twitching, without muscular inco-ordination. If the term chorea is to be restricted to the characteristic form of the child's affection, this case would not be an example of it. Yet in its occurrence during pregnancy, partial control by the will, the state of the heart, and complete recovery, its near alliance to chorea is obvious. Its mode of distribution pointing to a definite seat of irritation is the more significant from being altogether exceptional in that affection. The case may be well contrasted with the one that follows with reference to the chorea of pregnancy (p. 34).]

CASE XV.—*Chorea in early pregnancy. Acute Mania.*

Florence B., aged 21, married, admitted March 8, 1893, for choreic movements that have now lasted two months and been worse for the last fortnight. The movements began in the left foot, went to left arm, and afterwards to the *right* side. Speech has been affected for four days; she has difficulty in walking, but can feed herself.

The movements were first noticed after fright from seeing a child choking. She was in St. George's Hospital with rheumatic fever three months ago, and there waited on a child with chorea. She is now two months pregnant with first child.

Mother has had rheumatic fever, brother chorea, and sisters tetanus.

An emotional girl, with violent movement, now most marked on right side. Some contortion of face.

Heart sounds normal.

On the fourth day from admission the patient became very emotional, weeping and occasionally vomiting. She got but little sleep, and bromide of potassium was given.

After a week's residence the limb movements became more marked

but the emotional attacks less. There was not much sleep, however, and on this account 30 grains each of bromide of potassium and chloral hydrate were given at bed-time. With this she obtained fair rest for three nights; but morphia injection (gr.  $\frac{1}{6}$ ) was less successful.

On the tenth day of admission the woman became very restless and noisy. It was noted that the choreic movements, on admission most marked on the *right*, were now chiefly on the *left* side (though upon her discharge the side chiefly affected was again the right).

The emotional attacks now became more frequent, and in the intervals she would sit quite quiet, with a melancholy aspect, and pose, taciturn and occasionally violent so as to need restraint. The proper choreic symptoms had now wholly disappeared, and given place to those of mental unsoundness.

It was necessary, therefore, after a fortnight's residence, to remove the patient to an asylum.

CASE XVI.—*Chorea with Hemiplegia. Manner of the Association.*

Caroline C., a domestic servant, 21 years of age, was admitted into hospital in a condition of grave prostration, with a temperature of 102°, drying tongue, and an extensive but superficial sore over the sacrum. When put to bed it was observed that the face was in violent and continual movement, the tongue likewise being very unsteady and protruded with difficulty. The patient was confused in mind and unable to give any connected account of herself. She answered questions, however, more or less pertinently, but with great difficulty of utterance of the choreic kind. She moaned and cried out so as to disturb the ward, and her arms were restlessly thrown about. There was extreme tenderness above the elbows, the girl crying out upon slight touch. The urine was of high specific gravity (1030), and contained at first a trace of albumen. Large bronchial râles were heard over the chest; the heart-sounds very feeble and indistinctly heard.

The account of the patient, derived chiefly from her mother, was shortly this: in childhood she had twice had chorea, severely affecting both face and limbs, and some months ago while in service, she was laid up with rheumatism, apparently acute. Getting better of this, in about a month she was sent home. After six weeks' holiday she returned to her place and had now to carry a sick mistress daily up and down stairs, a service which she felt to be beyond her strength. She next began to suffer leg pains, and complained of weakness of the knees. A

month before her admission she travelled with her employers from Brighton to London. After this journey, and while sitting at tea, she suddenly lost all power in the legs, and was carried to bed. The history at this period becomes obscure, the patient having little recollection of the time. It appeared, however, from the account of the woman who had attended her, that she had complained of pains in the limbs, with cough and restlessness, and had shown much agitation of the muscles of the face.

The further progress of the patient need only be noted in so far as it concerns the relationship between the chorea and the paralysis. After two days' residence (during which, although not actively delirious, the patient was continually crying out, answering away from the purpose, and passing motions in bed, the bowels at the time being very loose) the extreme restlessness of the face became less. There now suddenly occurred a paralysis of the left orbicularis palpebrarum, and with this a complete lax paralysis of the right arm. There was no alteration in sensation, the early tenderness of the arms having disappeared. The tongue was still unsteady, the pupils equal and active. Temperature  $102.8^{\circ}$ ; pulse 102. The aspect and posture of the patient still indicated great bodily weakness. The bronchial signs remained; respiration between 35 and 40. The heart-sounds becoming more audible, a loud whirring systolic murmur was heard at the base, having certain peculiarities as to place which need not here be dwelt on.

After a week of this condition, during which diarrhoea continued at intervals, no control was exercised over bladder or rectum, and (with an improved mental condition) the prostration was extreme, the patient began to mend. The arm slowly recovered power, but remained for some time obviously weaker than the other. The orbicular paralysis also disappeared by degrees. The face and tongue, as has been mentioned, began to get steady when the palsy appeared, but it is to be observed of the tongue that it suffered a distinct relapse lasting but a few hours. On the tenth day after admission all signs of paralysis had disappeared, and the patient's natural manner, which was quiet and unexcitable, had returned. She was still very weak, with temperature over  $100^{\circ}$ , and pulse 120. The bronchitis continued, and there were some special physical signs at the left apex indicative of commencing phthisis. With the final disappearance, however, of the nervous symptoms, and the progress of recovery so far that the girl was able to get up during a part of the day, the case comes to an end for the purpose for which it is related.

[The relationship of the chorea to the paralysis in this case needs some analysis before attributing the two to the same cause.

1. *The chorea anticipates the paralysis.* By how long a time it precedes it is not quite clear. Having already had chorea twice, it is not remarkable that, with the incentive of overwork, and in convalescence from acute rheumatism, the same affection should return. It may be that both chorea and arm paralysis point to one and the same cerebral lesion. Yet the order of events must be kept in view, and it is this: facial chorea gives the first warning of cerebral change, incoherence of mind, pyrexia, and general prostration announce its actual advent; presently arm paralysis appears, which taken together with the valve disease and rheumatic history, contributes something to prove the lesion to be embolic.

2. *This chorea takes the form of a general and extreme restlessness* affecting all the face muscles alike. This is first noticed while the patient is still sensible, and not acutely ill; and, having stopped once, it is suddenly revived again for a short time, and then finally ceases. Of itself, therefore, this symptom would in no way help to determine the place of the supposed brain lesion. With paralysis of the right arm arising from embolus or extravasation, we should have expected rather a paralysis, or spasm, of the right lower facial region. We notice instead the gradual subsidence of an already existing facial chorea (the occurrence of which we may well accept in virtue of the patient's early history and previous attacks), with a paralysis of the *upper* part of the face on the *left* side.

Let it be assumed that we have in this instance some anatomical change in the corpus striatum or in the neighbouring convolutions which are in fact 'the same centre raised to a higher power representing the same muscles in more intricate combinations;' let it be embolism of minute vessels, or thrombosis, or any other pathological event calculated to produce an immediate trophic change. When did that particular event occur? In what relation does it stand to the chorea? This girl is ill and weak, and feels herself overworked, her legs fail, she has twice already had chorea, and now for the third time the face is found to be restless. Yet so little does this functional disorder affect her that she manages to travel in attendance upon her employers a journey of fifty miles. Reaching home she suddenly loses power, her face becomes more restless, and soon she is compelled to keep her bed altogether.

When admitted into the hospital the patient is pyrexia, delirious, and in a state of dangerous prostration; she presently becomes paralysed in the right arm and upper part of the face on the left side. Now, on the supposition of embolism or extravasation, or any like event, it is to these later symptoms and to this period that such event must be

referred, and not to the chorea which preceded them all. This latter was a revival of former choræas, happening under circumstances well calculated so to recall it. And not only is the functional disorder anterior to the proper cerebral symptoms, it fails to correspond with them in regard to its seat, it is not limited to a particular group of muscles but distributed over the face. It is a return of that which the girl had already suffered twice, and which any other kind of illness might have revived as well.]

CASE XVII.—*Repeated Chorea finally becoming permanent, with persistent contraction of flexor muscles of right hand and wrist. General Anæsthesia.*

Ellen K., a well-nourished, fair-haired girl, of quiet demeanour, was first admitted to the Westminster Hospital, Sept. 26, 1887, when seventeen, with chorea of right arm and leg. When eleven years old she was in the Evelina Hospital for the same disorder, and improved sufficiently in six weeks to be able to use the right hand. Her brother at the same time had chorea, as was supposed from fright, but he soon recovered. The patient having been discharged well, soon relapsed and went next to Guy's, where she remained a month. She again improved, but now began to drag the right leg. She was in Guy's a second time for three months—having in the interval had *several separate attacks from which she recovered completely*. From this time she began to get steadily worse. On admission to the Westminster Hospital, as above, the movements affected both sides, but chiefly the right arm and leg. The girl was quite unable to feed herself. The face was not affected, but there was some jerking of the head. She was perfectly natural and composed in manner; there was no cardiac murmur, no history of rheumatism or of fright. Careful inquiry from the mother elicited no more than that the father was 'rheumatic.'

When admitted, there was nothing to distinguish this case from ordinary chorea, but towards the end of her stay of four months, not only did she fail to improve, but it was observed that the movement became somewhat more repeating than at the first and tending to be rhythmical.

A note made at the time runs:—The movements are continuous, of moderate severity, and more or less rhythmical, the right arm and the legs being chiefly concerned, the left arm is nearly free from movement; the face and tongue quite steady. She can feed herself slowly and with difficulty. Pulse is regular 80, heart in all respects normal, except that its action is slightly uneven. Urine sp. gr. 1014 acid, without albumen.

Again, Oct. 15. Movements are much increased, extending to head, neck, and left arm, and incessant except in sleep. The deterioration is attributed to admission of a second patient with chorea. There is now distinct rhythm, *e.g.*, the right hand is repeatedly slapped upon the left and then withdrawn, while the knees are flexed with more or less regularity and with such energy that it requires considerable force to arrest the flexure movement.

The patient is now unable to feed herself, but she retains her cheerfulness, and is quite without emotion.

Common sensibility and that to pain are absent in the lower extremities, and much dulled over abdomen, chest, and face; knee jerks unequal; there is no ankle clonus. Pulse continues regular. No cardiac bruit.

The urea discharge was several times estimated. It varied from 1.4 to 2.7 per cent.

Arsenic was given over a long period, increased up to  $\eta$  viii. of the solution, and continued until the conjunctiva became affected. It produced no change.

A nightly dose of hydrate of chloral and bromide of potassium, of each ten grains, served to procure sleep. Daily shower baths apparently did harm, though the girl did not object to them.

With the patient's own concurrence a sheet was fastened round the body, securing and to some extent steadying the arms. Further, two arm pockets were attached to the sides of the bed, so placed that the girl could easily insert either hand into one or the other to restrain movement. The feet were also fastened by tapes to bottom of the bed. The leg movements were thus reduced, and the patient (who throughout bore her affliction with serenity) expressed her relief from this restraint. It was observable that with the greatest limb movement the body as a whole shifted but little as regards its position in bed.

For some time before her final discharge, at the end of 1888, the girl was completely anæsthetic over the whole surface excepting tongue and mouth; the right fingers and wrist had begun to contract, the *flexed position being maintained during sleep.*

When last seen, April 26, 1893, this young woman still exhibited arm and head movement of a jerky and repeating kind, not precisely resembling those of true chorea such as she had shown at the first. She was quite unable to feed herself or make any use of her hands, but walked with a short shuffling gait when supported on one side. The right hand was now firmly clenched and the wrist as firmly flexed, this position continuing, as the mother stated, during sleep. There was still,



so far as could be tested, complete anæsthesia of the surface excepting the tongue. The girl showed no mental deterioration whatever, and no sign of excitement or depression. The mother stated that she had been losing flesh for the last six months, but there was no evidence of organic disease. The heart sounds were natural.

[That the condition above described, though undoubtedly past cure in its latest stage, was in fact chorea there is reason to believe, not only from its answering to the definition but also from repeated recovery. And it is observable that so soon as it reached its chronic stage there was superadded anæsthesia and a permanent flexure of the hand and wrist persisting during sleep.]

CASE XVIII.—*Chorea in Middle Life occasioned by mental shock.*

James S., aged 38, a wire-worker, intelligent and unemotional, with no history of chorea, syphilis, or rheumatism, either personal or in his family. He had enjoyed good health up to three years ago, when he lost a child by death. He then became unsteady in the left arm, and soon in left leg and left side of face. Eighteen months ago, on the death of his wife, the chorea became general. He was for two months in St. Thomas's Hospital, and afterwards at Guy's. In the latter hospital he had an attack of acute illness described as rheumatism, and lasting two months. On recovery the limb movements became somewhat less.

The patient is in good general health, and least unsteady when employing his hands holding a book or turning over leaves. He is even able for a while to work at his trade. The choreic affection occupies the limbs, face, and speech muscles. In the limbs it is seen most in movements of extension, giving to his gait as he walks a curious spring for each step. The heart's action is regular except that with some beats there is marked prolongation of the first sound. When lying in bed the movements are almost confined to the upper limbs. The pulse is 94, temperature habitually subnormal, a little over 97°, rarely reaching 98°.

The patient remained in hospital for about two months, and improved unquestionably in his command of speech and power of walking. His own estimate, however, as to the measure of his recovery exceeded that of other observers. His poverty and precarious mode of living previous to admission made him so far a favourable subject for treatment, and such progress as he made towards recovery was due no doubt to improved nutrition from better living. As already

mentioned, he was always best when his hands were employed, and when discharged he believed himself well enough to resume his employment.

[This case affords an example of the very rare occurrence of chorea to a man of middle life, and not excitable or notably feeble in intellect, from the operations of causes similar to those which produce the disease in childhood. Each of his two attacks followed immediately on family bereavement. It is the more remarkable as he had not suffered chorea in early life. As regards rheumatism, the patient was certainly without it until after the chorea had become confirmed and general. The exact nature of the attack in Guy's Hospital described as rheumatism could not be made out owing to the man's infirmity of speech. The degree of intelligence through all the difficulties in the way of expression could not be accurately estimated, and there was no evidence as to his condition in this respect when first attacked. He was fairly educated, and fond of reading.]

## APPENDIX.

## TABLE A.

132 CONSECUTIVE CASES OF CHOREA, SHOWING (1) AGE AND SEX, (2) PREVIOUS ATTACKS, (3) RHEUMATIC CONNECTION, (4) EXCITING CAUSES, (5) STATE OF HEART, (6) LIMBS FIRST AFFECTED, &c.

## 1. AGE AND SEX.

Total number, 132. Boys, 33; girls, 99; viz.,  
 Of twelve years old: 2 girls.  
 Ten and eleven years old: 10 boys, 34 girls.  
 Nine years old: 7 boys, 20 girls.  
 Eight years and under: 16 boys, 43 girls.

## 2. PREVIOUS ATTACKS OF CHOREA.

2nd attacks, 27.  
 3rd     ,,     10.  
 4th     ,,     3.

## 3. RHEUMATIC CONNECTION.

132 cases give 7 who have had rheumatic fever;  
 14 (or 15) who have had pains, probably rheumatic;  
 6 that were doubtful;  
 5 not ascertained.

In 73 cases (the number in which that point is noted), rheumatism is in the *family history* of 19; of these 19, 9 have themselves had rheumatism, 10 have not. Of these 9 who have rheumatism, both themselves and in their families, rheumatism might have been the *immediate* cause of chorea in 2, and rheumatic fever in 1.

[The above is as exact an account of the rheumatic connection as the facts admit of; precise accuracy cannot be insisted on. See Chap. II., p. 40.]

## 4. EXCITING CAUSE.

Of the 132 cases :—

Supposed <i>exciting</i> cause not entered in .. .. .	20
Not to be ascertained in .. .. .	45
'Rheumatism' <i>exciting</i> cause in .. .. .	4 (? 3)
'Rheumatic fever' <i>exciting</i> cause in .. .. .	2 (? 1)
Fright or sudden shock in .. .. .	39 (? 2)
Some nervous strain, school discipline, hard lessons, &c., in	18 (? 2)
Previous illness in .. .. .	4

[The figures within the brackets denote proportion of doubtful cases under each heading.]

Hence it appears that of the 112 cases of which the exciting cause is presumably known, 57 or 53 (*say one half*) have their origin in some notable nervous shock.

## 5. IMPLICATION OF THE HEART.

Of the 132 cases :—

In 2 cases (of twelve years old) there was heart defect in none.

In 44 cases (of ten and eleven years old) there was heart defect in 16 (murmur in 13, irregularity in 3).

In 27 cases (of nine years old) there was heart defect in 3 (murmur in 2, irregularity in 1.)

In 59 cases (of eight years and under) there was heart defect in 26 (murmur in 16, irregularity in 10).

Hence, in 132 cases there is heart defect in 45, *the largest proportion of such defect appearing with the youngest children.*

## 6. PLACE OF ORIGIN OF THE CHOREA AS REGARDS THE SEVERAL GROUPS OF MUSCLES.

Of the 132 cases :—

No report as to place of origin in .. .. .	24
Chorea 'general' from the first in .. .. .	36
Chorea commenced in a particular group of muscles (whether or not becoming general after) in .. .. .	..from 69 to 72

Locality of first onset distributed as follows :—

Right side .. .. .	13 times.
Left side ... .. .	15 ,,

('Side' signifying generally arm and leg, but sometimes the arm only.)

Both hands .. .. .	10 (or 11) times
,, arms .. .. .	twice
,, legs .. .. .	once

Right hand	..	..	..	..	10 times
Left hand..	..	..	..	..	4 ,,
Right arm	..	..	..	..	3 ,,
Left arm ..	..	..	..	..	6 ,,
,, leg ..	..	..	..	..	once
Right leg ..	..	..	..	..	0
Face	..	..	..	..	4 times (?)

Hence, taking sides of the body (whether arm, leg, or both), chorea was at starting *right-sided* in 26, *left-sided* in 26; taking upper limbs against lower, it commenced in *the upper* in 36 (*hand or hands* in 25, *arm or arms* in 11), *the lower* in 2.

---

### TABLE B.

#### 45 CONSECUTIVE CASES OF CHOREA ILLUSTRATING THE SAME POINTS AS TABLE A, BUT IN SOMEWHAT OLDER SUBJECTS.

##### 1. AGE AND SEX.

Total number, 45. Boys, 10; girls, 35.  
 Over thirteen years: 3 boys, 7 girls.  
 Twelve and thirteen years: 2 boys, 12 girls.  
 Ten and eleven years: 2 boys, 9 girls.  
 Nine years old: 1 boy, 3 girls.  
 Eight years old and under: 2 boys, 4 girls.

##### 2. PREVIOUS ATTACKS OF CHOREA.

Second attacks: 11.  
 Several previous attacks: 1.

##### 3. RHEUMATIC CONNECTION.

Had had 'rheumatic fever,' 4.  
 Had had 'rheumatism,' 5.  
 Were doubtful, 11,

[As in the former table no more exact account obtainable.]

##### 4. EXCITING CAUSE.

Rheumatism (as *exciting* cause) in 2  
 Fright, or nervous shock, in 29  
 Over-study, 3; after scarlatina, 1; hysteria, 1  
 Exciting cause, not known in 9

## 5. IMPLICATION OF THE HEART.

Of the 10 cases over thirteen years old there was heart defect in 4			
Of 14 cases twelve and thirteen years old	„	„	4
Of 11 cases ten and eleven years old	„	„	4
Of 4 cases nine years old	„	„	1
Of 6 cases eight years old and under	„	„	3

Hence, in 45 cases there is heart defect in 16.

## 6. PLACE OF ORIGIN OF THE CHOREA AS REGARDS THE SEVERAL GROUPS OF MUSCLES.

Of the 45 cases :—

No report as to place of origin in 2.

Chorea general from the first in 5.

„ commenced in a particular group of muscles in 38  
as follows:—

Right side .. .. 4	Left hand .. .. 1
Left side .. .. 4	Right arm .. .. 1
Both hands .. .. 8	Left arm .. .. 4
„ arms .. .. 2	„ leg .. .. 1
„ legs .. .. 1	Right leg .. .. 1
Right hand .. .. 4	Face .. .. 5

Two were examples of spasmodic twitching of head and shoulders.

## RESULTS FROM THE TWO TABLES TAKEN TOGETHER.

## 1. AGE AND SEX.

Total number, 177. Boys, 43; Girls, 134.

Over thirteen years: 3 boys, 7 girls.

Twelve and thirteen years: 2 boys, 14 girls.

Ten and eleven years: 12 boys, 43 girls.

Nine years old: 8 boys, 23 girls.

Eight years old and under: 18 boys, 47 girls.

## 2. PREVIOUS ATTACKS OF CHOREA.

Second attacks: 38 (more than one-fifth).

Third attacks: 10.

Fourth attacks: 4.

## 3. RHEUMATIC CONNECTION.

Had had 'rheumatic fever,' 11.

Had had pains, probably rheumatic, 20.

Were doubtful, 17.

Not ascertained, 5.

## 4. EXCITING CAUSE.

Rheumatism, acute or not (as *exciting* cause) in 6.

Previous illness, exciting cause in 5.

Fright or some nervous shock or strain, exciting cause in 86, *i.e.*, nearly half (4 of these doubtful).

Overwork, 3: hysteria, 1.

Exciting cause not to be ascertained, 56.

„ „ not entered, 20.

## 5. PLACE OF ORIGIN AS REGARDS THE SEVERAL GROUPS OF MUSCLES.

Of the 177 cases:—

Place of origin not entered or not known in 26.

Chorea general from the first in 41.

Commenced in particular group of muscles in 110.

Locality of first onset distributed as follows:—

Right side .. .. 17	Left hand .. .. 5
Left side .. .. 19	Right arm .. .. 4
Both hands .. 18 (or 19)	Left arm .. .. 10
„ arms .. .. 4	„ leg .. .. 2
„ legs .. .. 2	Right leg .. .. 1
Right hand .. .. 14	Face.. .. .. 9

Five cases indefinite or anomalous.

Hence, in 151 cases (*i.e.*, excluding 26 not known):

Chorea was at starting *both* sided in 65 (or 66).

„ „ *one* sided in 72.

„ „ in the face in 9.<sup>1</sup>

„ „ indefinite or anomalous in 5.

It commenced in the *upper limbs* in 55.

„ „ the *lower limbs* in 5.

„ „ the *right hand* 14 times, against the *left hand* 5 times; but the *right arm* only 4 times, against the *left* 10 times.

Of all parts of the body, *chorea* began most often in *both hands*; while in either one or both hands it began in 38 instances, *i.e.*, in more than a quarter of the total number whose place of commencement was known.

<sup>1</sup> The numbers for the face are perhaps an under estimate, a slight degree of chorea of the features sometimes passing unnoticed.

## CASES OF DEATH IN CONNECTION WITH

In these 10 cases chorea was present so near to the time of death as to mention in the post-mortem record. Only the two last, however, are illustrations of death *by* chorea. The rest are examples of rheumatic heart disease, and in them chorea was not severe. Of the two fatal cases, one, Edith K., was under the care of Dr. Lees at the Hospital for Sick Children, the other, Thomas D., was under

POST-MORTEM APPEARANCE.					
No. & Vol.	Name and Title.	Pericardium.	Endo-Cardium.	Hyper-trophy.	Dilatation.
156 VII.	Winifred B. 8 years. Chorea-Carditis.	Distended with fluid. Blood stained lymph. Shaggy cords.	Fine and new vegetations on mitral.		
254 VII.	Charles H. G. 8 years 3 months. Chorea-Pericarditis.	Firmly adherent, also to chest wall.	Vegetations mitral and aortic.	+	+
58 VIII.	Louisa C. 8 years. Chorea-Pericarditis.	Universally and firmly adherent.	Fringe of small vegetations, mitral and aortic.		
197 VIII.	Joseph W. 9 years. Mitral Dis. Chorea.	Loosely adherent flakes of lymph. Right auricle lymph covered.	Mitral stenosis aortic valves natural.		
198 VIII.	Ellen S. 7 years. Rheumatic Endo-Pericarditis-Chorea.	Pericard partly adherent.	Endocarditis, but valves competent.	+	+
224 VIII.	John B. 7 years. Rheumatic Carditis. Chorea.	Universal firm adhesion.	Mitral and aortic vegetations (new).	+	+
138 IX.	Amy A. 8 years 6 months. Rheumatism-Chorea. Pericard. Morbus Cordis-Diphtheria.	Thickened, partly adherent. 1 oz. fluid.	Vegetations.	+	
139 IX.	Clara C. 6 years 6 months. Pericarditis-Morbus Cordis Chorea.	Firmly adherent.	Valves thickened not stenosed, endocardium healthy.	+	
98 X.	Edith K. 10 years 6 months. Chorea-Endocarditis.	Pericard natural.	Recent thin red line of vegetations mitral, rest healthy. See page 72 (note).	—	—
292 VI.	Thomas D. 15 years. Chorea-Endocarditis. (Westminster Hosp.)	Pericard natural.	Ring of small recent vegetations on edge of mitral.	—	—



OREA IN SIX YEARS (1886-92). TABLE C.

Donkin at the Westminster Hospital. They are entered here by permission of those physicians. Of Dr. Donkin's case it will be observed (as of so many that are fatal) that the supposed cause was alarm. I have no history of the other.

The titles are in all cases copied from the post-mortem books.

CLINICAL SIGNS.

Rheumatism.	Chorea.	Peri-cardial friction.	Physical Signs.	Nodules.
History of fright. No distinct history of rheumatism.	Slight.	Loud, double rub up to Death.	Aortic regurg.	0
1st attack 7 weeks ago.	Twitching of hands.	Pericardial friction.	Thrill systolic bruit.	+
Pain in right hand and wrist.	Not described.	Friction 14 days before Death.	Mitral systolic thrill.	+
Yes.	Not described.	No signs of friction reduplicated 1st at base.	Systolic and presystolic.	0
Yes.	Some chorea of lips.	No friction.	Systolic and presystolic.	+
Yes.	Not described.	Distinct pericardial friction.	Double mitral.	0
Yes.	Not described.	Double thrill.	Double apex murmur.	0
3 attacks.	Not described.	Friction.	Double apex murmur	0
0	Violent chorea hyperpyrexia.	—	—	0
0	Violent chorea after seeing a man killed.	—	—	0





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