THE HEART SYMPTOMS OF CHOREA.

BY OCTAVIUS STURGES, M.D., F.R.C.P.

Physician to the Westminster Hospital.

It is now generally admitted that there are certain heart symptoms proper to chorea apart from those which are due to its association with acute rheumatism or with anæmia. It is admitted as well that cardiac apex murmur is, more often than was at first believed, the sign of a functional disorder on the part of the valvular apparatus; the precise nature of this defective action has been in great measure elucidated, and it has been repeatedly shown both in chorea and other conditions that such murmurs are temporary and innocuous. With this double advance of knowledge, both in regard to the natural history of the disease in question, and the pathology of the heart, we may consider the cardiac symptoms of chorea from a new standpoint, with a clearer and more just apprehension than heretofore of the facts involved, and fuller and more definite material for their discussion. In what is to follow I propose to investigate the present state of opinion as to the nature and significance of the cardiac phenomena of chorea; to place such opinion side by side with the actual facts of the case; and upon this review, to consider whether by means of any existing hypothesis, or of any modification or combination of hypotheses, the question at issue finds reasonable solution.

The heart symptom of chorea chiefly discussed amongst us is systolic murmur. In regard to the rate and rhythm of the heart, as well as the precise seat of the bruit, there are, as we shall see, doubts and differences; but all observers allow that a soft systolic murmur is apt to arise in the course of chorea, independently of rheumatism or of anæmia, which is variable in tone, disappearing and reappearing, often influenced by

posture and by exercise, most audible in the majority of instances at the left apex, productive of no sensation of its own, and indicated by no outward sign of disturbed circulation.¹ Between this cardiac murmur and the choreic restlessness there is no equality or parallelism. No particular manner or degree of mismovement, and no particular temperament of patient is especially liable to this heart affection. The choreic murmur comes and goes secretly and without notice, there is no recognisable law of its occurrence, and when auscultation has discovered it, neither treatment nor prognosis is greatly influenced by the discovery.

But while these symptoms on the part of the heart, in their physical characters, limited duration and freedom from injurious consequences, remind us most of the so-called functional murmurs, yet have they a near relationship to substantive disease. This is not matter of inference, but of direct observation. There is no fact of chorea better established than this: that in the majority of fatal cases—dying either in the course of chorea or shortly afterwards—a fibrinous bead-like fringe is found edging the mitral valve, and occasionally the aortic valve also.² That this condition is the effect rather than the cause of the chorea, we are compelled to assume, in the numerous instances where murmur has gradually developed in the course of the muscular disorder. There are others, however, and especially those which arise out of acute rheumatism (be they few or many), where the sequence is different, and the cardiac murmur, from immediately preceding the chorea, may be taken to indicate an endocarditis which is certainly not the consequence and may possibly be the direct cause of the chorea.

But although endocarditis, upon this showing, seems to be connected with chorea in a double relationship, both as its cause and its product, there are obvious difficulties in the way of admitting it at all. For if endocarditis is really at the origin of chorea, why do the physical signs which should

¹ Such is the general description. Dr. Walshe, however, insists upon the regularity of action, and Dr. George Balfour asserts that the murmur "while it lasts, is constant and unchanging." See Walshe, 'Diseases of Heart,' p. 89; Balfour, 'Diseases of Heart,' p. 166.

² 'Statistics of Fatal Chorea.' 'Lancet,' July 17, 1880. 'Pathology of Chorea.' Dickinson, 'Med. Chir. Trans.,' vol. lix.

announce it so often lag behind, and why are they in themselves so equivocal and without the usual accompaniments and results of valve-inflammation? If, on the contrary, endocarditis is not the origin of chorea, but arises out of it, by what sort of agency is this accomplished, and why does not chorea, in virtue of this association, lay the foundation of organic heart disease as acute rheumatism does? Two conflicting doctrines are thus offered for our acceptance, neither of which we can wholly accept, and neither wholly reject. It is certain on the one hand that cases of chorea which happen to die (whether from the severity of the disorder or otherwise) very commonly exhibit a fringing of recent lymph on the mitral valve; it is not less certain on the other that the non-fatal cases (that is of course the vast majority) very rarely exhibit valvular disease in later life. It is certain that both the auscultation signs, and the subsequent history of choreic murmur correspond in most respects with the dynamic murmur of young women; it is not less certain that chorea prefers children rather than young women, and shows after death the actual material of endocarditis.

Such are the difficulties which meet us at the outset in endeavouring to explain the cardiac symptoms of chorea, and it is obvious that the account I have just given in regard to these symptoms does not contain the key to the solution. There still remain, however, other factors of the problem, some which are differently interpreted by different observers, and some which perhaps have not as yet received their due share of notice. Thus, for example, there is the character of the heart's rhythm, which some report to be irregular, and others undisturbed; and there is the further question as to the mode of incidence of cardiac phenomena in chorea in respect of age. It may be that the discrepancies which appear at present in regard to the precise characters and relative frequency of heart disturbance will disappear when we come to compare patients of similar age. We know, for instance, that the liability to such disturbance is in no direct relation to the violence or the particular method of the choreic attack; but we do not know that it has no direct relation to the age of the patient. Again, we know that choreic murmur may occur in-

dependently of any marked change in the cardiac rhythm; but we do not know that this is so at all periods of life, or that disturbed rhythm is not the rule rather than the exception in young children.

In what follows I propose to consider the characters and the frequency of cardiac disturbance in young children as contrasted with those who are older; to compare actual observations upon these points with the more general statements of authors; and, by the help of this comparison and the material it will require, to consider what hypothesis serves best to reconcile phenomena which at first sight seem to be conflicting. I take for this purpose the following:—

1. Dr. Dickinson's seventy-one cases (not 70 as stated) appended to his paper in the fifty-ninth volume of the "Medico-Chirurgical Transactions," and referring to children treated at the Great Ormond Street Hospital.

2. One hundred and thirty-two cases of children treated by myself at the same hospital, but at a late stage of their disorder.

3. Thirty-nine cases of older children and young adults under my charge at the Westminster Hospital.

4. Fifty cases similar to the last recorded by Dr. Owen in the ninth volume of the 'St. George's Hospital Reports.'

These four groups represent respectively, as will be seen more particularly in the sequel—

(1.) Young children from 2 to 12 observed at an early period of their disorder.

(2.) Young children of similar age observed at a later period of their disorder.

(3.) Older children with a few adults.

(4.) Older children, with a considerable proportion of young adults.

In Dr. Dickinson's table the number has to be reduced to 69, on account of two cases with the ages omitted. The children are all young. There is in fact but one, a girl, as old as 13, and but 3 as old as 12; the youngest is 3, the next youngest are 2 of 5; the ages most largely represented are 10 and 11. Taking the 69 cases, with a view to ascertain the incidence of heart affection at different ages, it will be found

that of the 21 children who are 8 years old and under, only 4 are free from heart affection, 3 of these being of the age of 8;¹ while of the 48 children above 8 years old, there are 17 free from heart disorder. If, however, we state the numbers somewhat differently, classing the children of 8 amongst the elder ones, we shall then find that in 11 children under 8, there is but one free from heart affection, while in 58 of 8 years old and over, there are at least 19 free from any abnormality of cardiac action which can fairly be reckoned.² As regards irregularity, the 69 cases give 20 such examples, 12 of irregularity alone and 8 of irregularity together with murmur. Of the 21 children 8 years old and under, there are 5 with hearts irregular only, and 3 with irregularity together with murmur. Of the 48 children over 8 years old, there are 7 with hearts irregular only, and 5 with irregularity together with murmur.

I take next a second group of children from the same hospital who were under my own charge, having been received from one or other of my colleagues.³ The ages are still between 2 and 12, as in the first group, but the children come under observation at a later stage of their disorder; the great majority were chronic cases, some few were convalescent. The total number of these patients was 132 (appearing as 137, from 5 of them having been twice admitted). The age distribution is as follows:—

12 years old and only	2
11 years old and	24
9 and 10 years old	47
8 years old and under, 36 of these being under 8	59
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¹ Of the 17 children of 8 and under having heart disturbance as many as 5 had had acute rheumatism, to which therefore the heart condition might be referred. On the other hand, of the 31 children over 8 having heart disturbance, as many as 6 had had previous attacks of chorea, to which period therefore rather than to the age recorded, the heart disturbance might be referred.

² I have made these calculations from the tables themselves (to which I refer the reader), and they do not precisely correspond with Dr. Dickinson's own summary (on p. 34 of his paper). This latter, however, has reference not to the *ages* of the patients, but to the several *causes* of the chorea.

³ I have referred to this series elsewhere, in connection with the rheumatic origin of chorea. 'Lancet,' Nov. 29, 1879. It is not given here *in extenso* out of regard to space.

In the whole 132, 47 (more than a third) have heart murmur or irregularity. The 59 children of 8 and under have 24 examples of heart disorder; the 72 children over 8 have 23 such examples. In other words, the proportion of heart disturbance is about 40 per cent. in the younger children, and about 32 per cent. in the older ones.

Again, if we take the extremes of age in both directions, that is to say, if we compare the 26 children of 11 and 12 (the oldest we have to do with) with 36 who are *under* 8, we have 9 of the former having heart disorder against 16 of the latter, the actual percentage of heart disturbance being thus somewhat greater in the younger than in the older children. Of cardiac irregularity, as apart from murmur, not including mere unevenness of rhythm, there are but 9 examples, 6 of these referring to the younger children.

These cases, therefore, representing the same ages as Dr. Dickinson's, but referring to a later stage of the affection, yield a much diminished proportion of heart disturbance of whatever kind. It is a little over a third, whereas in the former table it is over two-thirds. And what is still more remarkable, irregular action of the heart, largely represented in Dr. Dickinson's list, hardly appears in mine, and is almost confined to the younger patients.

It thus appears probable, taking these two tables as they stand, and comparing them together, first, that the heart affection of chorea is not less but more marked in early than in later childhood; secondly, that observation of the disorder at a late period discovers less heart disturbance than at an earlier, and especially that cardiac irregularity, a distinguishing feature of the chorea of early childhood, is found at the beginning rather than at the end of the attack.

Such, I say, are the conclusions to be drawn from a bare enumeration, and so long as we confine ourselves to children the majority of whom are not over 10, they are conclusions which need no correction. But when we enlarge the view and admit older children and adults; in other words, when we go from children's hospitals to general hospitals, we can no longer accept the mere figures without some attempt at adjustment on account of disparity of age. Thus, for example, all other

things being equal, the greater the age the larger the proportion of heart disturbance independent altogether of chorea. We have to consider besides, that in comparing childhood with adolescence, in respect of the signs we are considering, we have to encounter with the later period those functional murmurs (variously named, and attaching especially to young women) which, although they be altogether distinct from chorea as such, are not in practice distinguishable from it. Add to this that the older the patient the greater the probability that the chorea is not the first, but the second or the third attack, and the heart disturbance therefore but a revival of what has occurred in childhood, according to the well-known rule of the disorder to repeat itself. Such cases illustrate in fact the liability of *early* childhood in respect of heart sympathy: they are taken to illustrate the liability of the more advanced age at which the patient comes under notice. Again, some account must be taken of the previous occurrence of acute rheumatism, which is likely to appear in larger proportion in the histories of the older patients than of the younger, and to contribute its share of heart murmurs which are not choreic but rheumatic. It would be quite unfair, I say, to attempt the comparison we are now about without making allowance for discrepancies of this kind; without providing, that is to say, for the fact that the younger the child the more likely the heart disturbance to be intrinsic, the older the child the more likely to be accidental or contributed. In any exact computation as to the age-incidence of heart disturbance in chorea, it would be necessary of course to exclude from the comparison anæmic and chlorotic girls, as well as those who in earlier childhood had had either chorea or acute rheumatism. In the case of such children as we have been considering, all of them young, the correction necessary on this account would not amount to much, and we need not stay to make it; but in the case of the older patients we are now coming to, it makes a very wide difference. It would be altogether delusive and misleading to place the old and the young side by side with the view to an even comparison in respect of heart sympathy in chorea. Numerical equality between the two classes, or anything near an equality, would

indicate, without doubt, that the heart was more often implicated in the chorea of the young than of the less young.

How the figures stand in these respects with older patients, and how large a correction is needed for such reasons as have been mentioned, will be seen from the examples of chorea which remain to be quoted, viz., the 39 of Westminster Hospital and the 50 of St. George's Hospital. In the 39 Westminster cases 20 are 12 years old or over, 5 of them being young women. In the 50 cases reported by Dr. Owen in the 'St. George's Hospital Reports,' vol. ix., of two following years, as many as 27 are 12 years old and over, 11 of these being young women and 6 young men. We have thus in the four series of cases now rehearsed a constant rise in the ages of the patients. Dr. Dickinson's are the youngest, and the St. George's cases are by much the oldest, the Westminster series standing midway, and my own Children's Hospital cases differing from Dr. Dickinson's more in the duration of attack than in age of subject.

The 39 Westminster cases are distributed as follows:—

18 years and over (2 being young women)	15
12 years old	5
11 years old	5
9 and 10 years old	9
8 years old and under	5
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Of the whole number, 13 would have to be excluded, 5 on account of acute rheumatism at some earlier period, and 8 on account of previous attacks of chorea. The remaining 26 are made up of 19 who have no cardiac disorder, and 7 who have either murmur or irregularity. Of the 19 who have their hearts free, 15 are over 11 years old; of the 7 who have cardiac murmur or irregularity, only 1 is over 11.

In the two tables of cases of St. George's Hospital there are 15 examples of heart disturbance in 50 cases, 10 of these 15 are found among the 27 cases over 12 years old; only 5 are found among the 23 cases under that age. Upon this showing, it would seem that cardiac symptoms in chorea are not more rare but much more common after 12 years old than before.

Upon examination, however, of the 10 individuals exhibiting heart murmur and over 12 years old, it appears that 4 of them have had previous attacks of chorea, 1 had pericarditis while in hospital, 1 developed murmur just after acute rheumatism, and 2 were weakly girls, aged 14 and 15 respectively. There are thus 8 of the 10 whose murmurs are accounted for apart from chorea altogether. We get but two properly choreic murmurs in 27 choreic patients of ages ranging from 13 to 18, as large a proportion, I am disposed to think, as will be commonly found at that period.

There is indeed considerable difficulty in fixing the time of life when the liability to heart disturbance in chorea finally ceases. The evidence already adduced shows, I think, that childhood is particularly liable to it, and that irregularity of rhythm distinguishes this earliest liability, whether with or without systolic murmur. It is also generally agreed that the rare chorea of advanced life has little connection with heart disturbance. But of the middle period our knowledge is far from precise. For it so happens that at the very time when, with the progress of development, the frequency of cardiac murmur in chorea is obviously lessening there comes a period of life (and especially with young women) when the precise significance of the symptoms we are discussing is hardly discoverable. Murmur may be choreic or (so-called) anæmic. It would seem impossible at such time to determine with any approach to certainty the proper liability of the heart in connection with chorea. We know only that so soon as this interval of obscurity has passed, and, with the completion of adult life, the indications furnished by the heart are again trustworthy, cardiac sympathy in chorea is hardly recognisable.

From the facts now adduced several conclusions seem to follow. Heart symptoms in chorea occur most frequently during a period which is included between 2 years of age and about 7 or 8. The age of greatest liability to choreic heart disturbance thus partially coincides with the age of greatest liability to chorea. Again, the abrupt decline in the liability to chorea, which occurs at about 15, corresponds with a similar decline—if it be not rather a cessation—of the dispo-

sition of the heart to share in the disorder of the voluntary muscles.

It has been observed already that irregular cardiac action, either alone or preceding or accompanying the murmur, is a pretty frequent feature in the younger children of Dr. Dickinson's table. In my own children of similar age, but in a later stage of chorea, it is much less common. In the Westminster cases, where the patients are older, it is also rare, and in the St. George's cases, older still, irregularity hardly occurs. This particular symptom, in the younger subjects, is often the single sign of the heart's affection. I believe, although I cannot show by tables, that there are very few little children, if there be any, in an early stage of chorea, who do not exhibit in some degree unevenness of cardiac rhythm. Such unevenness will sometimes develop into murmur and sometimes not, murmur which is almost always variable with position, sometimes best heard over the pulmonary cartilage rather than at the apex,¹ apt to come and go, and often finally lost sight of before the chorea itself has disappeared. The second pulmonary sound is seldom (Dr. Walshe says never) either accentuated or doubled.

These statistics in regard to the age-incidence of the heart symptoms of chorea enable us to render a fuller account than that first given of the particular phenomena. Dr. Walshe's assertion that the cardiac action is regular must be limited to the older subjects, or to the later stages of the disorder. Dr. Bristowe's statement that the greater number of choreic patients have some cardiac defect, irregularity amongst the rest, must be qualified by adding that this large proportion refers to children, and that irregularity as a sign of heart disturbance is almost confined to them.

¹ The fact of mitral regurgitation being most audible in this situation, is well explained by Naunyn, as reported by Rosenstein (Ziemssen's 'Cyclopæd.' vol. vi. p. 122.) He points out that the second intercostal space, close to the left edge of the sternum, coincides with the point of the left auricular appendix, which winds round the pulmonary artery and lies in front of it. Now with the abnormal current of blood flowing towards the auricle, we can easily understand, says Rosenstein, "how the sound should be conducted to the spot mentioned better than towards the apex, more especially in cases where the appendix is long enough to lay its point close against the anterior wall of the thorax."

Thus the several factors of the problem with which we have to deal summarily stated are these:—

1. In the course of the chorea of childhood the heart is apt to become irregular or uneven, and its first sound to be followed by apex murmur, which is variable in pitch, influenced by posture, seldom audible in the axilla or at the angle of the scapula, and which disappears along with or shortly after the chorea, the heart and the circulation suffering no injury.

2. This liability on the part of the heart to what, from its signs, would seem to be a functional disturbance is independent of the violence or method of the chorea, but dependent upon the age of the patient, the younger children being the most, and the elder the least liable, while beyond childhood there is little if any liability of the kind.

3. These heart signs of chorea—acute rheumatism being excluded—give rise, as a general rule, to no symptoms whatever affecting the health or comfort of the child. They make no apparent difference to the prospects of recovery or the structural integrity of the heart. Nevertheless, choreic children having this murmur and happening to die, either with or shortly after recovery from chorea, very commonly exhibit a beading of recent lymph on the mitral valve.

Such, I say, are the chief statements which statistics seem to warrant. I will venture to add another, which, so far as I know, has never been statistically reckoned, but which no one will gainsay. It is, indeed, the most constant of all the heart symptoms of chorea, and met with at a later age than the rest. I mean the acceleration of the heart and pulse.¹

Now there are two hypotheses of the heart phenomena of

¹ Ziemssen says "cases of arrhythmia are certainly very rare, and there is hardly anything published about it." On the other hand, he regards acceleration of the action of the heart as common, and acceleration of the pulse as quite constant, although, as he believes (I am convinced erroneously) the bodily heat is not changed even in severe cases. (Ziemssen's 'Cyclopad.' vol. xiv. art. Chorea, p. 486.)

The heart symptoms mentioned in the text are not the only ones to be met with in chorea. I have heard presystolic murmur, and both accentuation and doubling of the second sound, and this in cases where there was sufficient evidence that the heart disturbance was purely choreic. But I am not able to say at present to what ages these exceptional signs chiefly attach, or what is their proportion to the commoner symptoms.

chorea, each of which has special regard to certain of the facts just mentioned, although neither is able to reckon with them all. The two are, in a sense, mutually antagonistic, inasmuch as what the one rejects the other prefers. Both theories have their supporters in this country. The one asserts that the heart symptoms of chorea are due to endocarditis, the precise relationship of the muscular disorder to the valve inflammation being variously interpreted. The other asserts that the cardiac signs are intrinsic, a part of the chorea itself. That there is some evidence in favour of each of these views we have already seen. The task before us is to reconcile them.

To the belief that the heart murmur of chorea is due to endocarditis I know of nothing in the actual clinical symptoms that can be opposed. Our information in regard to the physical signs of endocarditis *per se* is, indeed,¹ vague enough, while the probability of its occurrence in chorea is directly favoured, as we have seen, by post-mortem evidence. The distinction between organic and functional heart murmurs, founded on hard and fast lines as to the transmission of the bruit, is of doubtful validity. "A well-pronounced functional murmur," says an excellent observer,² "may be as diffused and transmitted as an organic." "A systolic apex murmur," says Dr. Bristowe, "is a positive proof of regurgitation." We have indeed, just as much evidence, so far as physical signs are concerned, of endocarditis in the course of chorea as we have of it in many instances of acute rheumatism. In both cases alike a soft and not conveyed murmur may be proved, in the event of death, to depend upon valve deposit, and in both

¹ "There are few diseases," says Rosenstein, "the presence of which is diagnosed so arbitrarily as that of original acute and subacute endocarditis." He proceeds to point out the necessity of "proving the existence of *other symptoms in addition to the mere murmur*, especially intensified second sound in the pulmonary artery, the localisation of the murmur, or transverse hypertrophy of the cardiac volume." Rosenstein, art. Endocarditis, Ziemssen's 'Cyclopæd.,' vol. vi. p. 90. Dr. Bristowe says, 'If in the progress of any of these diseases of which endocarditis is a common complication, we detect a recent cardiac murmur, and if further observation proves this to be a permanent phenomenon, we cannot reasonably doubt that endocarditis is present.' (Bristowe, 'Theory and Practice of Medicine,' 2nd ed. p. 522.)

² Dr. Nixon in 'Dublin Journal of Med. Science,' vol. iv. p. 575.

alike (I do not say to a like extent), when death does not so intervene, the murmur has been found to disappear. In frank recognition of these facts, Dr. Wilks expresses the belief that all mitral systolic murmurs associated with chorea are organic; and Dr. Sansom,¹ in a short summary of the prevailing opinions upon the subject, admirable for its conciseness, arrives at a like conclusion.

But when this much has been granted and endocarditis is admitted, how much are we the better? What is the mode of origin and what is the sequel of such endocarditis? It is but exchanging one difficulty for another. For we have to suppose, upon this hypothesis, not only that endocarditis may arise in and out of chorea, and that the younger the child the greater the probability of this event, but also that cardiac irregularity sometimes precedes and sometimes takes the place of regurgitation, whilst none of the injurious after consequences which attend endocarditis in its other relations are found to ensue here. In face of such grave objections, we are forced to admit that, although physical signs do not contradict it and post-mortem evidence may seem to be directly in its favour, the theory of choreic murmur (not to speak of the other signs) which invokes endocarditis is too difficult. It is inconsistent with all that we know of that inflammation in its various pathological relations; and apart from these we know remarkably little about it.

It is in fact by virtue of this vague knowledge that endocarditis maintains its hold in this connection. A form of heart inflammation special to chorea, and having a history and mode of termination of its own, is a creation of too shadowy a kind to be easily dealt with. But so soon as anything definite and within reach of investigation is alleged in regard to it, when, for instance, it is said (as Trousseau, Bouillaud, Roger, and other French authors say) that the endocarditis of chorea is in fact rheumatic, although the rheumatic element is itself latent, we have a distinct yet still but partially tangible issue. Of the relationship between chorea and rheumatism I do not propose to speak in this place. I have endeavoured to show elsewhere that although real it is rare and exceptional, and

¹ Sansom, 'Diseases of the Heart,' p. 89.

both Walshe and Hayden are of the same opinion. However this may be, no one will refuse to admit that the cardiac symptoms of chorea are observable sometimes when there is no open rheumatism either past or present. Now if in all such instances we are to say, in the first place, rheumatism is actually present, but after a secret and ineffable manner, and in the second place endocarditis is the consequence of that rheumatism, but it is an endocarditis which is exceptional in commencing after the chorea has begun and disappearing along with it, we account for the facts no doubt, but after a fashion which rather begs the question than provides for it.

We are thus compelled to regard the heart symptoms of chorea, taking them in their entirety and not by arbitrary selection—the early irregularity, the variable mitral murmur, the very frequent acceleration of heart and pulse with frequent fluctuations in the rate of both—as being altogether special and peculiar. They are signs of the heart's sympathy with the voluntary muscles, and are seen most at that early time of life when the antecedence of acute rheumatism is the least probable. The heart suffers by whatever mechanism or nervous influence, part and parcel with the rest of the muscular system, or rather it is apt to suffer. Such a conclusion, I say, in some sense or other, seems self-evident, and is indeed involved in the statement that the heart symptoms of chorea are met with in no other association.

Obvious and inevitable as it may be, it is far from being easy of application. For if it be granted, in the words of Dr. Walshe, "that the apex murmur of chorea is plausibly ascribable to disordered action of the muscular apparatus connected with the valve," why does not the same disordered action extend to the ventricular walls? It may be answered (although Dr. Walshe does not admit as much) that in young children there is both disorder of rhythm and regurgitation; but the answer is insufficient. These two signs do not commonly concur nor do they vary together. And it is not young children alone that we have to consider, but the elder patients as well; and in these latter, while murmur is not infrequent, any marked irregularity of cardiac rhythm is

certainly rare. Thus we have 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," says Dr. Hayden, "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 acts 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."¹

There are other objections equally formidable. It is not in the nature of chorea to dis sever the normal combinations of muscular movement. It stirs the very same muscles that are employed by purpose or emotion, and moves them in a similar way, only less precisely and with less definition. Apart from anatomical considerations, therefore, a choreic spasm of the heart, which should select the papillary muscles and leave the cardiac walls, would be untrue to the pattern of chorea elsewhere. And again, if this choreic conduct of the heart be but an extension of the same disorder which affects the other muscles (an extension, as Dr. Kirkes points out, to which there is no parallel in any other involuntary muscular organ), the most violent and general chorea ought to exhibit this cardiac disturbance the most. But it is not so. On the contrary, the violent chorea of puberty has seldom either cardiac murmur or cardiac irregularity, while the chorea of the young child, which is but rarely violent and is sometimes rather a paresis than a disorder of movement, exhibits both these forms of heart affection very often.

And even if these difficulties should be overcome or the ground of some of them disputed, it would still, I think, be hard to understand how the great centre of circulation and (in one sense, at all events, within the experience of everybody) of emotion should become the seat of a mis-movement at all resembling the choreic contortions of the visible muscles without giving rise to sensations of grave disquiet, sensations, be it remembered, which, so far as young children are con-

¹ 'Diseases of Heart and Aorta,' p. 268.

cerned, the favourite subjects of such cardiac disorder, are conspicuously absent.

It would thus appear that both the hypothesis which supposes endocarditis to be the cause of choreic murmur and the hypothesis which supposes an extension of the muscular infirmity to the heart to be the cause of it are fallacious. Both theories commit us to too much. And if, in this strait, we have recourse to vaguer expressions having regard to the "impoverishment" or "vitiation" of the blood, we neither provide an adequate explanation nor allege a demonstrable cause. But while thus compelled to abandon one position after another, we must beware lest by too precipitate a retreat we quit ground which it is essential to hold. We may even venture so far upon the assured facts of the case as to make some sort of stand and to affirm something. If the views just announced cannot be held in their entirety they must be held with some modification or other. There is no alternative. Part of the perplexity arises, as has been shown, from a consideration of the harmlessness, variability and limited duration of the choreic murmur. But if these facts are in themselves secure we may rely upon them to force at least this conclusion, that the conduct of the heart in chorea is similar to its conduct in anæmia. Whatever explanation, therefore, applies best to the precise mechanism of the so-called functional or dynamic murmur cannot be altogether foreign to the circumstances of chorea.

It is now generally admitted that the murmurs once called hæmic, anæmic, and by similar names implying dependence on some blood change, are much more often than was at first supposed produced by actual regurgitation, due to a temporary defective action on the part of the mitral valve. Such murmurs are to be heard in purpura, in anæmia, in the general muscular relaxation after various nervous strains, in epilepsy, as well as in typhus and enteric fever, and even in simple pyrexia. This dynamic mitral bruit may, by exception, be harsh and far sounding, audible in the axilla and even at the lower angle of the scapula, yet nevertheless by its ultimate disappearance give ample proof that whatever may befall the valve during its continuance (and there is some reason to believe that a material change actually occurs) is not abiding

or permanently mischievous. So much, I think, is generally admitted. The question for us now is, whether the same explanation of the mechanism of this murmur which applies to its association with the conditions just mentioned applies also to its association with chorea.

No one, so far as I know, has more ably discussed this subject than Dr. Nixon.¹ His exposition of the various ways in which dynamic apex murmur may arise, and of "the perfect unity and correspondence in action of the fibres of the walls of the ventricles and of those which are connected with the valves, which is needed in order to ensure complete closure," suggests to him the inquiry whether this nice correspondence of movement might not be so disturbed by altered nutrition or innervation as to render such closure for a while defective. The tremor of the voluntary muscles, which is apt to follow upon their violent exercise, is adduced as being analogous to this disordered cardiac action. Dr. Nixon concludes that functional mitral murmur results from "this want of correspondence between the fibres of the ventricle, which obliterate the cavity, and those which close the valve;" an altered function due, as he believes, "to some defect in the vital power or condition of the heart itself, which leads either to atony of the papillary muscles or derangement in the rhythm of their movement." In the further observations of the same author as to the means of separating this murmur from the organic, he alludes to its changeable character, alteration or even disappearance with the upright posture, and the absence (in most instances) of accentuated or double second sound. Thus, in all its essential features, the murmur which Dr. Nixon describes corresponds with that of chorea; and, except in cases of temporary debility, anæmia and certain conditions already particularised commonest in the female sex, it is seen in no other connection.

Dr. Hayden, assenting in part to these views, would attribute the dynamic murmur not so much to a want of correspondence between muscular fibres, as to atony or parietal debility of the walls of the left ventricle.² Weakness and relaxation of the papillary muscles, he admits as a possible cause of mitral regur-

¹ 'Dublin Journal of Medical Science,' vol. lv. p. 572. &c.

² Hayden, loc. cit. p. 274.

gitation. Spasm, or irregular action of the same muscles, he does not admit as a possible cause of it. This regurgitant murmur, "caused by atony and partial yielding of the walls of the left ventricle at the acme of systole," Dr. Hayden has long associated with anæmia, purpura, and excessive use of tobacco. His later observations lead him to include chorea and fatty heart in the same category.

But the latest and least reserved expression of the opinion that dynamic murmur is in fact dependent on muscular paresis is that of Immermann.¹ "The muscular tissue of the heart," says he, "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 any undue exertion of the cardiac muscle a temporary paresis of the muscoli 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. As the organ regains its normal energy these grow fainter and ultimately disappear." He goes on to describe the fatty degeneration of the papillary muscles which sometimes ensues upon simple anæmia, and which explains both the occasional permanence of these murmurs and their occurrence in connection with fatty heart.

It will be admitted, I think, that both the time of appearance and precise manner of the choreic murmur, as well as its mode of origin and duration, varying intensity and limitation to the period of youth, are all nicely provided for in the hypotheses just rehearsed. If only it be admitted that the heart's disorder is really a sympathy with the muscular disturbance, an inexact and precarious correspondence between the ventricular contraction and that of the papillary muscles would seem to serve for chorea even more than for the conditions to which Dr. Nixon applies it. Still better, and without making any assumption as to the heart's sympathy, might we admit the doctrine of partial paresis of the valve apparatus, and insist that the excessive muscular movement of chorea is the precise condition which, upon the hypothesis, should give rise to

¹ Immermann, art. "Anæmia," Ziemssen's 'Cyclopæd.' vol. xvi. p. 399.

murmur such as we hear in token of fatigue on the part of the heart. Adopting either of these views, we might say with Ziemssen that "change in the valvular sound is *à priori* more likely in chorea than in any other affection."¹

But the matter is not so easily settled. Let it be granted that the choreic murmur is due to a functional disorder on the part of the mitral and probably also of the tricuspid valves, by means of which a temporary and variable regurgitation is permitted (and I think so much of admission is inevitable), we have to ask, in the next place, why this regurgitation, whether it be due to faulty adaptation of the several parts of the heart to one another, or to the paresis of fatigue, is not in harmony with the other symptoms? If the murmur betokens a sympathy on the part of the heart with the disordered action of the other muscles, it ought to appear when chorea is the most generalised; if, on the other hand, it betokens a fatigue paresis, it ought to appear when the chorea is most violent and prolonged. But it does neither. The sympathy of the heart (if it be rightly so called) goes not with the manner or the degree of the chorea, but, as had been shown, with the age of the patient. And, moreover, we should be taking a very partial view of the subject were we to limit the heart symptoms to cardiac murmur. This, as we have seen, is but one, and not even the chief, symptom of choreic heart disturbance. There is besides the unevenness or irregularity which belongs especially to early childhood, and, most constant of all, there is the accelerated pace of the heart. Again, we have not merely to account for the presence of these additional signs, we have to fit them into their places, and to show why the heart, seldom failing to share the muscular disorder in one way or another, exhibits this mode of disturbance or that according to the age of the subject. We may accept either account of the mechanism of dynamic murmur, and without much violence apply it to suit the circumstances of chorea: that is a little gain, no doubt. But when that is done there remain these further symptoms, which, unlike valvular murmur, can compare with nothing to be found in other analogous conditions, unless indeed the cardiac unevenness and its acceleration remind us of what is sometimes met with in

¹ Loc. cit. p. 440.

hypochondriacs and hysterical women, the least promising subjects to throw light upon obscure places in pathology.

It would seem then that to understand the conduct of the heart in chorea we must look to chorea itself and the modifications it exhibits at various ages. Here is an affection which both in absolute frequency and in its tendency to implicate the heart, attaches itself to young children most of all, to older children somewhat less, to adults hardly at all. At each of these periods chorea, both in the limbs and at the heart, shows itself after a particular manner. If, then, the action of the heart in chorea is throughout in real harmony with that of the voluntary muscles, the clue to the secret we are in search of must be sought in the special characteristics of those periods of life to which chorea especially attaches—characteristics, that is to say, in which childhood and youth differ from infancy on the one hand and adult life on the other. If the reader is not already wearied by the length of this exordium and the somewhat circuitous path we have had to pursue, I would venture in such space as remains to take a final survey of the subject from this point of view.

I would observe, in the first place, that the chorea of the post-infantile period is the immediate successor of the muscular spasm or convulsion of the infantile period. At an age varying with the "forwardness" of the child, the one liability takes the place of the other. The very same material cause (as for example, intestinal irritation, or the troubles of dentition) will excite convulsion in infancy, and chorea a little later. Again, the chorea of early childhood will sometimes commence with convulsion, or the two affections may alternate, the same limbs being affected in both cases. Little, of course, can be certainly known as to the actual sensations of the child at this early age, or of the mode in which physical pain gets mentally interpreted; but it depends probably in some measure upon the degree of completeness of such interpretation whether the response shall be by way of convulsion or by way of chorea. However this may be, the general muscular restlessness of the young child which passes for chorea is not only in close union with spasm in the way just mentioned, it often bears a direct resemblance to it in form. There is a uni-

versal unsteadiness in which the muscles of the abdomen and of respiration share as well as the diaphragm and larynx. This generalised chorea which thus succeeds to convulsions is, so to speak, the simplest form of the disorder, without variety so far as the movements of the several parts of the body are concerned, and without any mental admixture. It is an aggravation of the natural unsteadiness of childhood, an universal and equable overmovement or mismovement extending to all those regions which it is the nature of chorea as a motor disorder to affect, and it is this merely. There is no incoordination, for the muscles have not yet learnt to act in concert; there is no distinction between the movement of one part and another, for the several muscular uses are not yet differentiated, and the limbs are stirred indifferently without method or purpose. Such general, featureless agitation of the body is the proper response to a source of irritation which appeals to no one part in particular and has no mental representation.

With maturer age the bodily movements become varied and definite, and each department of the muscular system sets about its separate business. And as with movement so with mis-movement. The chorea of later life is a definite deformity which alters and disfigures the natural carriage in this way or that according to its seat. It selects those muscles which are the readiest to respond to emotion, and moves them as they are wont to be moved by the passions and employments of life. Thus the choreic distortion of the face is one thing, of the legs another, of the arms another; and this separateness becomes the more distinct as the individual grows and his individuality, as it is called, becomes more and more marked. The mismovement of chorea betrays the age of its subject, no less than does the natural movement. And as life goes on chorea becomes more and more distinctly localised. In its latest form it is a partial disobedience or misconduct of certain muscles, rendered inveterate by habit, and incorporated as it were with the rest of the bodily movements.

My conjecture is that the conduct of the heart in chorea is in strict accordance with these successive modes or differentiations of the muscular disorder. The heart sympathises with

chorea in one way or another throughout life, or at any rate until that period is reached when the affection is both rare and difficult to identify, and has ceased to apply to the whole system. Not to speak for a moment of the valvular murmur, the heart is unrhythmical or irregular in early childhood,¹ accelerated in youth and left undisturbed in old age. Thus the earliest period of heart sympathy, that of irregularity, with or without acceleration, corresponds with chorea, a purely motor disorder, the earliest transition from infantile convulsion and differing from it mainly in the character of the movement and the implication of the emotional centres; but resembling it in its exciting cause, wide range, and conformity everywhere to a single pattern. In the heart as well as the limbs, in the tongue, the speech, the diaphragm, in all the parts in fact accessible to emotion, yet without any mental representation of emotion, the choreic disturbance of little children is a motor disorder, merely excited very often, like the earlier convulsion, by a material irritation.

In like manner the cardiac acceleration of youthful chorea, with or without murmur, corresponds with that form and period of the affection when the several muscular movements have been differentiated, and there is no longer a single and uniform response, but each department of the body liable to chorea responds after its own manner to a disorder which is now first mentally apprehended and intimately connected with emotional exaltation. As to the limbs there is no longer a mere restlessness impartially distributed, but complicated disorder of movement in which paresis and incoordination are variously blended, and which select by well-marked preference those muscles which are the readiest exponents of emotion. And as to the heart, in place of disturbed action and rhythm there is the proper emotional response with which everyone is familiar in his own person, an acceleration, namely, and often knocking impulse as well.

¹ 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.

Now at both the periods we are now considering, in childhood and in youth, both with cardiac irregularity and cardiac acceleration, we have, or we may have, apex murmur. It has been shown already that for such murmur no explanation is applicable except the functional one. And indeed, in the case of choreic patients, at that time of life when the so-called anæmic murmur is common, I know not how it is to be determined whether valve defect is due to chorea or to anæmia. Perhaps the same causes may operate in both cases, and the explanation of Dr. Nixon of faulty adjustment of valve and ventricle may serve for both. But, apart from these, there are the children to be considered. And, if we are still to apply the argument that the heart symptoms of chorea are in constant harmony with the rest, it is necessary to remember that there is another element in chorea besides those that have been alluded to. I mean *paresis*.

The real character of this choreic paresis has been somewhat obscured, I venture to think, by the prominence that has been given to those examples of it which have chanced to affect one side of the body only, and thus to give promise of material for illustrating the morbid anatomy of chorea. I would only remind the reader in this place as pertinent to the present inquiry, that a strictly localised muscular paresis very often mixes with choreic restlessness in children; that it is often very sudden both in its access and departure, giving to the affection an aspect of deformity which of itself it has not; that it may be recognised especially in a droop of the wrist or a falling of one shoulder, or feebleness of grasp of one hand; that it selects this part or that according to no known rule, and may in some instances show so prominently as to give the patient the appearance of paralysis rather than of chorea. Now all that can be said of choreic paresis as we see it in the limbs may be said as well of choreic murmur as we hear it at the heart. Both are characteristic of childish or at least youthful chorea, both are sudden in their development, variable in degree, and apt to come and go without any other corresponding change in the character of the disorder.

If then weakness and relaxation of the papillary muscles be

admitted as a possible cause of mitral regurgitation, and if a debility of this kind may arise temporarily from tobacco smoking or nervous exhaustion, much more should it arise in chorea where, apart from muscular fatigue, there is a special liability to paresis extending, as it would seem, to all the muscles which the disease is able to affect. And observe that this relaxation on the part of the papillary muscles, which is common both to anæmia and chorea, and productive of regurgitation in both, is not of the same origin in the two cases, nor always accompanied by precisely the same signs. In anæmia it gives rise to an equable murmur which only ceases with the gradual recovery of the patient; but in chorea it is (or it often is) a variable and inconstant murmur which hardly repeats itself precisely for two consecutive beats, and which appears and disappears without apparent cause, as does the similar paresis of the voluntary muscles, with which indeed, as I have repeatedly noticed, it may be strictly synchronous both in its time of arrival and duration. Moreover, with a murmur having this origin, we are rid of the difficulty which attaches to the doctrine of a fatigue paresis, and would require that the valvular defect should be in direct relation to the duration and severity of the chorea, which would associate it therefore, not so much with childhood, as with that violent form of chorea which is commonest at a later period of life.

Yet still the old objection will recur. Whence comes it that a temporary paresis of the valve apparatus is found to be associated after death, if not with endocarditis, at all events with a deposit of recent lymph on the edges of the mitral valve? That this appearance does not represent a true inflammation of the endocardium is rendered probable by its bead-like arrangement and limitation to the margin rather than the auricular surface of the valve. There is never ulceration, and where time has been allowed to elapse between the chorea and death, the valve is found to be in a normal condition. Only in cases where death supervenes either in the course of chorea or very shortly afterwards is this appearance met with, and in these not always. There is ample clinical and post-mortem evidence that it is neither lasting

nor injurious.¹ Mindful of this strict limitation of the facts to be interpreted, and of the necessity in the last resort of reaching a solution upon the principle of exclusion, the question suggests itself whether an imperfect closure of the mitral orifice, or rather such uncertain action of the valve segments as closes it at one time but not at another, might not, at the approach of death, and when with the flagging action of the heart fibrine is very readily deposited, so determine the seat and method of such deposit, as to produce at the edge of the valve the bead-like appearance that we see; the condition which immediately precedes death being one of the essential factors of the occurrence.²

It may be said, indeed, that whatever is true of the dynamic murmur of chorea must apply equally to the same murmur in anæmia and elsewhere. But of the post-mortem appearances of simple anæmia we know absolutely nothing. There is, however, a disease having a striking likeness to chorea in respect of its preference for the female sex, near connection with hysteria and with mental emotion, and apparently anomalous blending of functional with organic disturbance. In exophthalmic goitre we have at first acceleration of the heart and afterwards mitral murmur, a murmur which sometimes wholly disappears with the complete recovery of the patient, and sometimes is the precursor of organic mitral disease.

In conclusion, I would briefly recapitulate the facts which

¹ I venture to extract the following from a paper of my own, already referred to, on 'The Statistics of Fatal Chorea':—

This post-mortem appearance occurs equally in those that die in connexion with chorea, and in those that die directly of it. Thus of Dr. Dickinson's 22, there are, as I reckon, 12 dying *with* but not *of* chorea; of these 10 have mitral valve vegetations and 2 have not. Of the 10 dying *of* chorea, 7 have vegetations and 3 have not. In Dr. Peacock's 3 fatal cases, all dying of chorea, 2 have vegetations and 1 has not. Out of 5 cases, the only examples I can find of fatal chorea with perfectly healthy heart, all save one, an old woman, are typical instances of death in and by chorea.

² The suggestion in the text is in fact that of Dr. Dickinson. "The beads," he writes, "are usually confined to the inner surface of the mitral valve and arranged along the attachment of the thin edge, where a line of minute but abrupt prominences is presented to retrograde blood, but an arrangement of more gradual slopes to blood flowing normally. Thus possibly the collection of fibrine is the consequence, not the cause, of the regurgitation." ('Pathology of Chorea,' loc. cit. p. 87.)

it has been the object of this paper to establish, together with the hypothesis which they seem to support. The facts are that in chorea the heart is apt to sympathise with the voluntary muscles at all ages up to the adult period, this sympathy being shown as well by dynamic murmur as by accelerated action, unevenness of rhythm, and, not seldom, the excited impulse common in hysteria, the particular manner of response being dependent upon the age of the patient. The hypothesis is that these several modes of heart affection correspond with as many modifications of chorea, which are exhibited not in the heart only, but in the voluntary muscles as well; these several regions sharing jointly, each in its own degree and after its own manner, in a disorder the area of whose influence is co-extensive with that of ordinary emotional disturbance: and, particularly that in all such variations the motor element of the affection is represented mainly by irregularity and unevenness of cardiac rhythm, the emotion element by acceleration, and the paresis element by dynamic murmur.