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CHOREA MINOR

WITH A TABLE OF FIFTY CASES

BY

A. CHARLES E. GRAY M.B.C.M.
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Appendix. Table of Cases of Chorea.
In the following paper I propose to describe Chorea Minor or Sydenham's Chorea as it occurs in children, giving a brief account of the characteristics and symptoms of the disease, and in particular such explanation of its causes and origin as seem to me to be warranted by a consideration of the statistics as to etiological factors and the observation of actual cases. My observations are drawn from a series of over fifty cases, under my exclusive care in private practice and in the out-patient department of Paddington Green Childrens Hospital, and in addition I have had access to the records of cases treated as in-patients in this hospital. All these are cases which have occurred in children under fifteen years of age. I do not intend to refer, except incidentally, to the manifestations of chorea in association with pregnancy, nor to those occurring in adults apart from pregnancy, more especially as in the case of the latter there is good reason to suppose that the nature of the disorder is essentially different.

History. The name Chorea Sancti Viti was originally applied to a dancing mania which prevailed in the 14th and 15th Centuries, on account of the fact that the Strasburg authorities ordered the sufferers to be taken to the Chapel of St Vitus at Zabern, to be cured by the influence of the Saint. The disease about to be described, differs entirely from this, but the term was applied to it by Sydenham, who wrote the first
description of the complaint in 1693. Since then the
disease has attracted widespread interest and has been
closely studied by numerous observers both on the Con-
tinent, in America and in this country.

Character and Symptoms. The essence of the
disease is the occurrence of constant, irregular, invol-
untary, purposeless movements, with apparent inco-ordin-
ation of, or rather interference with, voluntary move-
ments, accompanied by mental alteration and occasionally
actual muscular weakness.

The first thing that strikes an observer, on seeing
an example of this disease, is that the patient is af-
fected by an extraordinary and abnormal restlessness.
He is unable to remain still for a moment. The face
is contorted by involuntary, ridiculous grimaces, while
the head is moved constantly from side to side; the
shoulders are twitched and hunched up; the fingers are
flexed and extended and intertwined one with another,
the arm at the same time being turned and twisted in
every direction; the feet are shifted restlessly about.
If the child is asked to show the tongue, it is pro-
truded with a jerk and as instantly withdrawn, or if
kept out with an effort, constant tremors may be seen
taking place in it. All the voluntary muscles of the
body are involved, those of the trunk as well as those
of the extremities, but in the milder cases it is the
arms and face which are chiefly affected, and in which
one most easily sees the manifestations of the disease.
With all this there is no true tremor of individual muscles; the twitchings are in reality ordinary movements and the abnormality consists in their involuntary character, their purposelessness and their frequency. The condition has been well described as an "incoherence of muscular movement". The interference with ordered movements is, as might be expected, very considerable. Thus in walking, the legs are thrown about in all directions, so that the gait is unsteady and staggering, and a fall is frequently the result. As involuntary relaxation of the muscles takes place as frequently as contraction, objects laid hold of are dropped, (which is one explanation of the complaint of paralysis sometimes made by the friends) and any action is performed jerkily and irregularly, both on account of the involuntary cessation of willed contractions and the supervention of unwilled movements. If the patient is able to write, the character of the disorder is well brought out, the pen is held with difficulty, the fingers constantly shifting their grasp, and when put to paper the regularity of the letters formed is interrupted by sudden excursions in a wrong direction. In severe cases, all voluntary acts become quite impossible; the child cannot stand, feed or dress himself, and flings himself about in bed so that, if not prevented, he falls out or injures himself against adjacent objects. In some cases an effort of the will serves to arrest the movements for a time, while in
others it only increases the unsteadiness. Most frequently the effort to hold one limb steady aggravates the movements in other parts, so that in mild cases, when the face is ordinarily in repose, the effort of holding out the tongue at once produces facial contortions.

Along with these symptoms one can hardly fail in the majority of cases to observe that some mental alteration is present. The expression is vacant and slightly dazed, response to questions is slow, memory when it can be tested is often found impaired, and the edge of mental faculties seems to be taken off. Speech is in a large number of cases affected, apparently both on account of the mental affection and the muscular difficulty in articulation. The weakness of muscles rarely if ever amounts to actual paralysis; the grasp is feeble or it may be weaker on one side than the other, but there are apparently some cases in which the movements are very slight or absent and the weakness of a group or groups of muscles the chief symptom. These cases may have begun with some twitching, the movements passing off and the weakness persisting.

The onset of the disease is occasionally sudden, so that from a normal state of health there is developed in a few hours a marked degree of movement, but more commonly the affection begins gradually and is present for some time before it is recognised. It follows from this that the first signs are misinter-
preted and the child may be punished for what is not his fault. A boy or girl usually quick and eager at school is unaccountably restless and fidgety, his attention is constantly wandering, his lessons are badly learned, and this goes on till the characteristic jerking appearing in the limbs warns the friends that the child is suffering from actual disease.

In a certain proportion of cases the movements are at first limited to one side of the body, and in a small percentage the disease remains throughout its course confined to one side, and we have a true Hemi-chorea. More commonly the disease, at first one-sided, spreads in a few days to the other side and becomes general. An interesting feature of this one-sided character is that the distribution of the movements is sometimes similar to the distribution of paralysis from central disease; the disorder, entirely one-sided as regards the arm, is observed to be more general in the face, and to affect slightly the opposite leg as well. On the question as to which is most frequently affected at the commencement of the disease the evidence is somewhat conflicting. Osler, Russell, Jackson and Money say that it begins most frequently on the right side, while Austin, Flint and Trousseau say the left. Gowers finds the numbers nearly equal for each side, 31 right 33 left. Of my own cases there were 11

1. Chorea.
in which the disease was observed to be at first, or throughout one-sided, the right being affected in five, the left in six. We may therefore reasonably conclude that it manifests no special predilection for either side.

With regard to the temperature it may be stated that, speaking generally, the disease is non-febrile in character. A survey of the temperature charts of a number of cases treated in hospital shows that the curve is frequently somewhat irregular, but, except when some intercurrent condition, such as an attack of tonsilitis or acute rheumatism exists, the temperature rarely rises above 100 degrees.

It will be well to consider the symptoms in detail, as they affect the various systems of the body.

The Alimentary System is not usually specially affected. In very severe cases the mouth becomes dry, the tongue thickly coated, the teeth covered with sordes and the appetite is entirely absent. The tongue is frequently bitten by the spasmodic closure of the jaws, resulting in some cases in extensive ulceration; and on account of the excessive movement the administration of food by the mouth becomes impossible. The organic reflexes are not affected however, and deglutition, when food can be taken by the mouth, is normal, and the functions of the rectum are not disturbed. In mild cases the appetite and digestion generally remain fairly good throughout.
As regards the Haemopoietic System, there is almost always a greater or less degree of Anaemia to be noted. It is rare however for this to be a very marked feature, though in some cases it is so. Considering the well known potency of rheumatism to produce anaemia, especially in children, it is probable that the rheumatism so frequently accompanying chorea is largely responsible for the anaemic condition, but no doubt the exhausting effects of the disease have also a share in producing it.

In connection with the Circulatory System, the commonest complication of chorea arises. In a very large proportion of all cases auscultation of the heart reveals the presence of a murmur or murmurs. The most frequent of these is a soft blowing murmur, audible in the mitral area, systolic in time, sometimes accompanying, sometimes replacing the first sound. Next in frequency comes a presystolic mitral murmur, either alone, or along with a systolic. Further, in some cases, murmurs are to be heard over the base of the heart. But besides these murmurs, which are certainly endocardial, there is to be heard, in a small percentage of cases, a to and fro exocardial rub, showing evidence of the presence of pericarditis. Generally speaking it is only on auscultation that any hint of cardiac disease is obtained; the pulse may be a little quickened, generally is, but the usual symptoms of cardiac disease in adults in the shape of dyspnoea or dropsy are very rarely present.
In a considerable number of cases, the endocardial murmur is noticed to be present when the patient first comes under observation, while in others, its development takes place during the subsequent course of the Chorea. Again in some cases the murmur disappears as the patient begins to recover, while in others, and these the large majority, the murmur persists after all choreic movements have entirely ceased.

Statistics as to the actual frequency with which endocardial murmurs are present, vary considerably according to different observers, which is no doubt partly to be explained by a similar variation in the importance attached to examination of the heart. One or two examinations are not sufficient to establish the non-existence of a murmur, as it may arise at any time during the course of the disease. Of 47 cases under my own care, a distinct Mitral Systolic murmur was present in 26; two of these having, in addition to the Systolic, a presystolic murmur. In three of the remaining 17 cases, the first sound in the mitral area was noted to be impure. In three of the 26 cases the murmur disappeared when the choreic movements ceased, but in all the other 23, it persisted as long as the patient was under observation. This gives a percentage of 60.46 in which a mitral systolic murmur was present, and the results of other enquirers who have given special attention to the heart, very closely approximate. The records of cases treated in the wards of Paddington Green Childrens Hospital during the six years 90-96 show that out of a
total of 63 cases 54 or 53.96% had a systolic bruit at the apex. Garrod found murmurs present in 56.25% and Gowers puts the number of cases with cardiac disease at something over one half. The statistics of the British Medical Association Collective Investigation Committee show, on the other hand, a somewhat smaller proportion namely 115 in 449 cases. More significant, however, is the result of an enquiry by Osler, into the subsequent history of Chorea patients. He carefully examined 140 cases at an interval of time after the attack varying from two to sixteen years, and ascertained the following facts— in 51 cases the was normal, in 17 cases there was disturbance of the heart, probably functional in character, and in 72 cases or 51.42% there were definite signs of organic heart disease.

Much discussion has arisen as to the significance to be attached to the presence of these cardiac murmurs. By some it is maintained that they are purely functional haemic bruits, owing their origin to the condition of anaemia produced by the disease, and evanescent in character. Sturges, who has devoted much study to the disease, has brought forward a theory that they are due to irregular action of the Chordae Tendineae, that in fact the muscular disorder extends to the heart. This must certainly be described as a pure hypothesis. There is not a trace of evidence in its fa-

1 Med.Chi.Trans.1889.Vol.72
2 Diseases of the Nervous System
3 Coll.Invest.Record.Vol.IV
4 Osler.Chorea
5 Chorea. 1895
your, and any irregularity in the action of the heart is most usually conspicuous by its absence, at any rate when the murmurs first arise. By others it is believed that, in the majority of instances, they are indicative of cardiac disease and are due to endocarditic changes in the valves. Of the correctness of this latter opinion there seems to me no doubt. Undoubtedly haemic bruits do arise and disappear in the course of chorea, but that, in the larger number of cases, the murmurs are due to endocarditis seems to me to be established by the following facts.

1. If the murmur were generally a haemic one we should expect it to be most frequently audible at the base of the heart, the situation in which anaemic murmurs usually develop, whereas in chorea it is the mitral orifice which is most commonly affected.

2. The murmur is not seldom present at the onset of the chorea before debility and anaemia have come on.

3. In the majority of cases the murmur persists after the termination of the chorea, and if the cases are followed up, definite organic heart disease, with its attendant train of symptoms is found to develop. I have already referred to the result of Osler's enquiry: Stephen Mackenzie following a somewhat similar line of investigation found that the murmurs were persistent in from 60 to 80%. Many interesting examples of the subsequent serious results of choreic endocarditis are recorded by Cheadle; but the case of R.A. a boy

of twelve under my care is fairly typical. When he first came under observation for an attack of chorea, a presystolic mitral murmur was found to be present. There was a history of rheumatic fever in near relations but the boy himself had had no rheumatism if we except some pains in the legs which occurred at intervals during the preceding few months and were considered by the friends to be "growing pains". Four months after the onset of the chorea and several weeks after its disappearance, in spite of the fact that he had been treated by absolute rest in bed, he was noted to have tumultuous and irregular action of the heart, slight dilatation and a well marked presystolic murmur, accompanied by a faint short systolic murmur. Two years later, the systolic murmur had become more pronounced, the presystolic was as before; the boy was breathless on the least exertion and quite incapacitated for any pursuit in life involving active physical exertion.

4. The endocarditis is sometimes accompanied by pericarditis, the organic nature of which is not disputed.

5. In fatal cases of chorea changes in the valves are found in the vast majority, which in no way differ from those present in cases of endocarditis associated with acute rheumatism. Fatal cases are so rare that the number of Post Mortem examinations is small, but of those recorded 93.75 per cent

1. Sturges.
showed definite endocarditic changes in the valves. Of sixteen post mortem examinations collected by Fagge he says "In every one of these there were fibrinous nodules found on the aortic or mitral valves. In only two of these was a history of rheumatic fever obtained and in seven its occurrence was explicitly denied."

These facts would seem to me to prove that in the majority of cases of chorea the heart is more or less affected and that the nature of the lesion is, in most instances, an actual inflammation of the endocardium. This endocarditis associated with chorea is, like the endocarditis of childhood generally, seldom severe. Symptoms referable to the state of the heart are rarely present and if the heart were not examined it might easily escape notice. On the other hand the disease is very liable to be progressive, successive slight attacks tending to occur, with the result that in a few years the heart becomes seriously and permanently crippled.

Though this mild character of the attack is the rule exceptions occur, and pericarditis with effusion, or rapidly progressive endocarditis, is responsible for the result in not a few of the fatal cases of chorea.

The Respiratory System. In a few cases the rhythm of respiration is disordered, the diaphragm and other muscles of respiration sharing in the spasmodic affection. The breathing may be gasping, or hurried, or jerky and irregular, a deep breath being followed by

shallow ones, or actual pauses occurring. In my experience this is not frequently present in any marked degree but slight irregularities similar to those observed in nervous persons are fairly often to be seen. Beyond this the respiratory system is not specially involved.

**Integumentary System.** In two of my cases an attack of herpes zoster of a typical kind developed during the course of the chorea. In both the eruption appeared on the right side of the chest. The attacks ran a normal course and pain, though present, was not a marked feature. In a third case, there was a herpetic eruption round the mouth. I have not met with any records tending to show that herpes is a common complication of chorea, (Osler found only two cases out of 410) and can only therefore conclude that the occurrence was accidental. Hutchinson and Nielson consider that arsenic produces herpes zoster and I may therefore mention that both the cases which developed herpes on the chest had been taking arsenic in ordinary doses, but in one this had been stopped for three weeks before the eruption appeared. Erythema Nodosum, Multiple Erythema and Purpuric Urticaria are said to occur rarely, but I have not met with any examples.

**The Urinary System** is not affected by the disease. The urine on examination does not as a rule

1. Loc. cit.
2. Jamieson, Skin Diseases.
contain albumen or any other abnormal constituent, unless we except a pigment called Urohaematoporphyrin. This pigment is very commonly found in the urine of patients suffering from rheumatic fever, so frequently indeed, that Dr. A. Garrod, who has investigated the question, states that he has detected it in the urine of nearly every case, in which he has looked for it. He also examined the urine of twenty cases of chorea and found it present in fourteen. In view of the fact that the pigment is not found in other diseases, this may regarded as one of the evidences of the connection between these two diseases.

**Nervous System.** I have not met with any alterations of common sensation, nor do abnormal sensations, such as tingling or numbness, seem to occur. It is a question whether the muscular sense is affected in any way or not, but there is certainly no marked derangement. In young children the difficulty of making accurate investigations into subjective conditions is naturally very considerable. The disease is of itself not painful, but it is not uncommon to meet with complaint of pains in the limbs during its course, flitting about and transitory in character, which are in all probability manifestations of the accompanying rheumatic state. The special senses of sight and taste and smell are unaffected. The ophthalmoscopic appearances are usually those of health, though slight optic neuritis has been described as occurring in rare cases.

As I have already incidentally mentioned the organic reflexes are normal. With regard to the condition of the tendon reflexes, much diversity of statement exists. Thus Henoch states that he has generally found the reflexes, especially the patellar reflex increased, while Osler, in a series of 50 cases examined, found them normal in 26, and diminished, or not elicited, in 24. As a matter of fact, I have found all conditions of the knee-jerk to be present in the same case at different times and although it is impossible, on account of the many exceptions which occur, to make any very positive statement, the condition, which I have most frequently observed, is, that at the commencement of the disease the knee-jerks are either diminished or absent and that as recovery takes place they slowly return to the normal; on the other hand I have notes of cases in which the knee-jerk was exaggerated or not elicited during the whole course of the chorea. Not infrequently the two knee-jerks are elicited unequally, and though, in some cases, the greater departure from the normal seemed to be on that side which was most affected by the movements, I have been unable to satisfy myself that any rule could be laid down with regard to this.

I have already described the characteristic affection of the voluntary muscles. The muscular weakness, which is a not infrequent symptom of the disease, is generally comparatively slight, and rarely if ever amounts to complete palsy. It most commonly affects

2. Loc. cit.
the arms, more rarely a leg, there may be a slight difference in the grasp in the two sides, nothing more. It may arise at any time during the chorea; may even precede the development of the twitching. Occasionally, as was seen in one or two of my cases, a slight weakness may persist for a time after the movements have ceased. In some cases, when the disease is more or less one-sided, the weakness is rather apparent than real; the patient finding attempts at voluntary movements in one arm so unsatisfactory, refuses as far as possible to make any use of the affected limb.

The electrical irritability of the muscles has been examined in cases where only one side was affected. In some cases there was no alteration in the reaction but in others a distinct increase of irritability, both in nerve and muscle, to both forms of current, was observed. A qualitative change in the response to the Voltaic current has also been described as occurring in some cases; the anodal closing contraction occurring as readily as the cathodal closing contraction, instead of requiring a stronger current.

**Coordination.** I do not consider that there is any real alteration in the coordination of voluntary muscular movements, apart from the interference with ordered movement produced by involuntary muscular contractions. What, in my opinion, takes place, is that an intended and perfectly coordinated movement is interfered with by secondary, unwilled, cortical dischar-

ges, starting in the motor centres, and reaching, it may be, the muscles taking part in the willed movement or only affecting others, with in either case, a like result, namely, that the action is performed jerkily, or arrested altogether. This is contrary to the expressed opinion of many, that incoordination is the essence of the disease. Thus Gowers states that voluntary movement is irregular partly from disturbing spasms, partly from an incoordination which is not always related in degree to the amount of spontaneous movement. In some cases the latter may be slight, while incoordination is great, and conversely voluntary movements may be almost steady, though there is much spontaneous movement. Now it appears to me that the disturbing spasm is the sole cause of the apparent incoordination and, that in the above statement, the effect of the will and of the attempt at voluntary action on the spontaneous movement is not sufficiently taken into account. Observation of actual cases brings out, I think, the following facts, 1st. That the involuntary movements are nearly always increased by attempts at voluntary movements. This increase is not necessarily in the limb which is performing the action, though sometimes it is. Thus a protruded tongue, which is slightly tremulous, becomes markedly more so if the child be directed to close the eyes tightly, and, if the hands are engaged, the restlessness of the feet and the contortions of the face be-

come much greater. In some cases this is even more marked, the patient while at rest being perfectly still and the twitching only appearing when any voluntary movements are made. 2nd. In the majority of the less severe cases, in which alone observations of this kind can properly be made, the patient is able for a short time to control the spasms sufficiently to execute an ordered movement. This control cannot be kept up for long; the inhibitory power apparently becomes tired out and the movements reappear. A good instance of this ability appears in the fact that most children who have learned to write can manage to sign their names even when much twitching is present. I have samples of writing by children whose hands were in a state of constant typical choreic movement when simply under observation and who yet contrived to write their names in a very creditable manner. It follows from this that the handwriting is no fair index of the amount of disorder present. The writing however shows very clearly the characteristics of choreic movement. The regular outline of the letters is suddenly interrupted by an excursion of the pen in a totally wrong direction, exactly similar to what would occur if the child's elbow were struck by a second person.

On the following page are samples of writing by choreic patients, which show fairly well the peculiarities mentioned above.
This capacity for inhibiting the movements is sometimes much more extensive. Thus, in the case of a girl Agnes R., treated as an in-patient in Paddington Green Childrens Hospital, it was noted that she was able when told to keep perfectly still, while when not consciously under direct observation there was very much movement. But indeed one of the difficulties in describing chorea and in defining the nature of the condition is that cases differ so much from one another; the knee-jerks absent in one case are exaggerated in another, inhibition of spasm a marked feature of a third is conspicuous by its absence in a fourth, so that, except for the disordered movements, there is hardly a single condition of the nervous system, exhibited by a particular case, which will not be found absent in another.

Mental Functions. In almost all cases some deviation from the normal mental condition is present. This is commonly comparatively slight and often fails to attract the notice of the patients' friends. The keenness of the intellectual faculties is dulled, the child appears a little stupid and has a vacant, listless expression, the power of fixing the attention, never very highly developed in a young child, is observed to be less than formerly, and memory is very frequently impaired. The difficulty in speech, often noticed, appears to be due not only to the muscular difficulty of articulation, but also to the mental inefficiency, to be in fact a slight condition of aphasia. These mental symptoms are sometimes present before the onset of the characteristic movements and their cause not being re-
cognised the child is not seldom punished at school for his want of attention. In addition to this there may be disturbance of the moral sense in various ways. The child's character is temporarily distinctly altered for the worse and he is described as irritable, perverse, bad-tempered and unlike himself. Often this condition is one of depression, and the patient is said to be low-spirited, to be constantly bursting into tears without adequate cause. In almost every case the child is abnormally easily excited and this state of excitement generally increases the spontaneous movements. This was well illustrated by one of my cases in which, during the later stages of the disease, the child was reported to be almost perfectly well at home, but as soon as she was brought to hospital, and especially when under observation, the muscular agitation became violent and constant. In one or two cases however the opposite effect was produced and the patient, described as constantly jerking the limbs while at home, was under observation comparatively quiet.

In rare cases the mental disturbance is very much more profound and becomes the most prominent feature. There may be a condition approaching maniacal delirium and delusions and hallucinations are occasionally present. None of my cases exhibited such a severe disturbance as this, but the child Ada D., during the height of her first attack, for which she was treated as an in-patient in hospital saw "people pursuing her"
and had other visual hallucinations on several occasions. Her second attack was much milder and the mental alteration was not marked.

The mental symptoms generally pass off along with the disappearance of the choreic movements, but they sometimes persist for long after all other signs have entirely ceased. This was noted in one of my cases, where impairment of memory and stupidity remained for some months after, but as it is almost invariably the case, entire recovery eventually took place.

Sleep is usually restless and disturbed. When the movements are very severe they may prevent natural sleep altogether, but sleeplessness is also due to the cerebral condition, as even when there is little movement there may be much loss of sleep.

In the vast majority of cases the movements cease entirely during sleep, but, in rare cases, it occasionally happens that they persist to a slight extent even in sleep.

Complications. Endocarditis and pericarditis, occurring in chorea have already been referred to, though it is a question whether they should really be regarded as complications. In some cases embolism has been known to occur. Acute articular rheumatism may arise during the course of the chorea, and attacks of tonsilitis are occasionally seen. Notwithstanding that the nature of the complaint suggests some likeness to epilepsy, convulsive seizures are rare in chorea. Attacks ap
parently of the nature of petit mal occurred in one case treated in Paddington Green Hospital, and cases in which epileptiform seizures occurred have been described by some authors.

As a rule intercurrent diseases have no definite effect upon the course of chorea. Henoch mentions cases in which pleurisy, measles and tonsilitis developed and ran their course during the attack of chorea, without any definite effect upon the latter. Similar observations were recorded in the cases of Emily H. who had measles, and Mary D. who had tonsilitis during the time they were under treatment for chorea in hospital.

The duration of the disease is extremely variable. The average is often stated to be about ten weeks. Very few cases last less than six weeks and many very much longer, three, four or even six months. A small percentage persist for more than six months, thus Gowers found. If cases at the National Hospital for Paralysis and Epilepsy in which the disease had lasted more than six months and seven more than a year, while in Angel Money's statistics there were ten cases which had lasted more than a year and five more than three years. Cases, however, of such very long duration, are many of them cases occurring in adults and probably belong to a different category, and, in estimating the average duration of chorea in children, should be left

1. Loc. cit.  2. Loc. cit.
out of account and considered separately.

There seems to be really no definite course for the disease; the condition once established tends to wear itself out in different times, in different individuals. Variations in observations too are, no doubt, introduced on account of the fact that it is difficult to say exactly when the disorder has come to an end. Long after the cessation of the obvious manifestations careful examination will in many cases reveal the presence of occasional slight twitches in the hands or face, or an emotional disturbance will give rise to an apparent but temporary recrudescence of the disease. In general, cases are discharged from hospital before complete cessation of the movements and outpatient cases often cease to attend when the parents no longer notice much disturbance. Hence it is probable that the real duration is often understated. In the cases which I was able to observe till complete cessation of all movement the average time was 15.08 weeks.

Some instances of long duration are doubtless cases in which relapse or recurrence has taken place, before the primary attack has completely subsided, some examples of which I have observed.

Generally speaking the more severe the attack the longer the disease lasts, but as with so many other facts connected with chorea, this is not invariably the case and no definite rule can be laid down.

Recurrence. Chorea has a marked tendency
to occur again in the same individual. Thus, of 410 cases recorded by Osler, 170 or 41.46 per cent had more than one attack; of these, 35 had three, 10 four, 12 five, and 5 six attacks. Of my cases 21 out of 46 had more than one attack. This gives a percentage of 45.65 of recurrences, but, as my observations only extend over a period of two years and are confined to children under fifteen, the real proportion is probably somewhat higher. Of Gowers cases one third had more than one attack, while one case had as many as nine. Obviously the figures must always be below the mark, as the subsequent histories of many patients treated in a first attack, especially those attending hospitals from which statistics are mostly derived, cannot be followed up.

There does not seem to be any common factor governing the incidence of these recurrences. The ratio of males to females is much the same in second attacks as in first, though after two attacks there is an immense preponderance in favour of females, and immediate exciting causes are not more frequently noted. The interval between two attacks is very variable and may be any time from a few days to a few months. It seldom exceeds two years. (I am not here considering the case of chorea gravidarum. Many of these cases might be regarded as instances of recurrence under special conditions, as there is a previous history of chorea in childhood in a large proportion.) Probably when the interval is very short it

1. Loc. cit. 2. see Chorea Gravidarum, Mo. Camn  
should more properly be regarded as a relapse than as a separate second attack. As might be expected, from what has already been said, organic heart disease is found more frequently in second than in first attacks. Gowers noted its presence in half the cases of second and third attacks, and in all the cases having more than three attacks. In the case of Lena G. however, in my series, the heart was noted in a fourth attack to be free from signs of organic heart disease.

As regards the character and symptoms of the disease there is no marked difference between recurrences and first attacks, but, though exceptions occur, the severity and the duration of subsequent attacks tend, in my experience, to be rather less than first.

Etiology. Age. Chorea is essentially a disease of childhood. Of 522 cases recorded by Osler, 509 occurred before the age of twenty. Of 80 cases recorded by Garrod and 69 by Heringham the average was 10.23 and 11.35 respectively. In my own collection gathered at Paddington Green Hospital, where the age limit of patients treated is 14 for girls and 12 for boys, the average age of 102 chorea cases was 7.91. To quote Gowers "nine tenths of all cases occur between five and twenty and four fifths between five and fifteen."

The disease is rare below the age of five. One case is recorded by Sturges in a girl of two years eleven months, and the earliest case I have found recorded is one in a boy of two years and six months, who was treated for a typical attack of chorea in Paddington Green.


Hospital last summer. It is also exceedingly rare above the age of twenty-five, and though cases do occur in later life, it is probable that they are of a different nature.

The average age incidence is somewhat earlier in boys than girls. Thus, in Garrod's cases, the average age of the males, at the first attack, was 8.91, that of the females 11.56, while of my cases it was 7.17 for boys and 8.2 for girls, the total number being 102. Of this number there were four cases under five (namely one and a half years, two just over three years and one over four years,) three of which were girls, but on the other hand, while there were only three cases among the boys, in a total of 29, there were 23 girls out of 73 which were ten years old or more. The disease is very rare in boys after sixteen and almost all the cases between twenty and thirty are furnished by women.

Sex. Chorea is enormously more common in females above than males. Of the mentioned number, which includes both the in-patients at Paddington Hospital and the cases treated by myself, I find there were 73 girls and 29 boys or 73.5 per cent of females. A combination of statistics quoted by Gowers gives 365 boys to 1000 girls or 73 per cent; while of Osler's cases, 390 in a total of 554 or 70 per cent, were females.

Station in Life. There is no evidence that the disease is more frequent in one class of society than several authors another, though state that it is more common among the poor. With regard to this statement

it must be remembered that statistics are chiefly gathered from hospital practice, which is practically confined to the poor, and that, of course, as the well-to-do classes number not more than one fourth of the whole population, the actual, though not the relative frequency, will as a result be smaller.

**Season.** The influence of the season on the onset of the disease does not, in this country, seem to be great, though it would appear that there is a slight falling off in the number of cases arising in July and August. Combining two sets of statistics, namely Money and Gowers, the numbers commencing in each quarter of the year were, First 102, Second 93, Third 53, Fourth 77. Of my cases the numbers were similarly, First 29, Second 22, Third 24, Fourth 36. In this connection it is interesting to note, that Lewis, in America, found a seasonal relationship to exist between chorea and rheumatism. Comparing the average number of attacks per month of chorea and rheumatism, it was found that the variations in the occurrence of chorea corresponded with those of rheumatism, but were uniformly a month later.

**Geographical Distribution.** The disease is more commonly met with in large towns than in country districts. Isambard Owen, from the results of an enquiry into the distribution of acute rheumatism, rickets, chorea etc., concludes that, while rheumatism is pretty nearly universal throughout the whole of the British Islands, "chorea follows to a very great extent the distribution of rickets and like it accumulates in the great indus-
Heredity Influences. It does not appear that chorea itself is frequently inherited. I found only 5 cases in 95 with a history of chorea in the father or mother's family. Angel Money found 14 in 214, Sturges 6 in 100, and the Collective Investigation Committee 14 per cent with a family history of chorea.

Though most writers agree in thinking that chorea occurs in neurotic families, statistics do not show very many instances of neuroses in the immediate families of choreic patients. Clouston states that he constantly meets with chorea in the children of his insane patients, in the children of dipsomaniacs and of epileptics. Nevertheless, the numbers given in published statistics are not large. Sturges found neuroses in 13 of the ancestors of 100 cases, viz.: insanity in six, epilepsy in one, mental distress at child's birth in the mothers of three, drunkenness in one and hysteria in one. Money in 214 cases found a history of epilepsy or fits in eight families, imbecility in one, convulsions in one, and delirium tremens in one; altogether eleven or 5.14 percent. Gowers puts the neurotic heredity at one sixth of all cases, but this is including a history of chorea itself. A certain allowance must be made for the difficulty of obtaining a history of the neuroses, but while in only two or three cases have I obtained on enquiry a history of

2. The Neuroses of Development, p. 64.
3. Chorea, 2nd Ed.
4. Loc. cit.
epilepsy or insanity, a considerable number testified to one or other of the parents having in a marked degree a nervous or emotional temperament.

There is another disease however, namely rheumatism, with which the hereditary relations of chorea are very close, a history of this being given in a much greater number of cases than of any other disease. Statistics on this head differ considerably, varying no doubt on account of the bias of the individual observer and the kind of evidence accepted as satisfactory.

In compiling statistics of rheumatic family histories it seems necessary to exclude every thing except an actual attack of rheumatic fever, the term rheumatism standing in the minds of the public to denote so many different conditions. It must be admitted however that, by doing so, we shall obtain results which in all probability fall below the truth, as a certain number of cases of rheumatic fever are only vaguely remembered by the friends as rheumatism.

Turning now to the actual statistics I find that in 23 cases out of 42 or 54.76 per cent there was a distinct history of rheumatic fever in immediate blood relations, while in several other cases there was a history of rheumatism. Garrod finds a history of rheumatic fever in 32.5 per cent, Heringham in 33.3 per cent and Gowers puts the rheumatic inheritance at 45 per cent. Cheadle finds a rheumatic history in the patient or relations in 75 per cent. These figures would not be of much val-

1. Loc. cit. 2. Rheumatic State in Childhood.
ue without control statistics as to the normal incidence of rheumatic fever. Fortunately investigations on this point have been made by one or two observers. Garrod and Cooke taking 500 patients at St. Bartholomew's Hospital, who were not suffering nor had suffered from any rheumatic affection or chorea, obtained a family history of acute rheumatism in 21 per cent or, if the patients suffering from tonsilitis be excluded, as possibly rheumatic, of 19.78 per cent. On the other hand in 100 patients who had rheumatic fever, a family history of the same disease was found in 35 per cent. Cheadle made careful enquiry into the family history, of 492 children admitted for diseases of all kinds into the Great Ormond Street Hospital for children. In 173 there was a clear history of acute rheumatism in immediate blood relations. Of these 173, 38 had developed unmistakeable rheumatic affection, equal to 20 per cent. Taking the remaining 319 in whom no history of joint affection in relatives could be traced, only 15 developed articular rheumatism, equal to 4.5 per cent.

An interesting feature of the hereditary association of chorea is that not unfrequently the two conditions, viz., neuroses and rheumatism occur together in the same family. The case of P. A. in my series is a good example of this. One of the mother's sisters suffered from rheumatic fever, another had hysteria with marked

1. Lancet. 1888. i. p. 110
2. Rheumatic State in Childhood. p. 27
ovarian symptoms, while the mother herself was alcoholic and after the birth of the child decidedly melancholic. The patient himself had rheumatic pains and endocarditis immediately before the onset of the chorea; so that we have here an excellent instance of the effect of the combined neurotic and rheumatic inheritance in producing chorea. In other cases the hereditary association of chorea with rheumatism shows itself in the development of the two diseases in different members of the same family, one child having acute rheumatism and another chorea. In one family under my notice, one child had chorea and another asthma.

Temperament. That temperament has much to do with the development of chorea there seems to be no doubt. The subjects of the disease are excitable, nervous, active brained children, as a rule. Out of 38 of my cases, in which the question was enquired into, there were 32, equal to 84.2 per cent, in which the child was described as specially nervous or excitable.

Previous Illnesses. There are only two diseases with which chorea is found closely associated namely rheumatism and scarlet fever, and the connection with the former is so marked that, as I hope to show, it may be regarded as a causal connection. This bearing of the question will be discussed more fully later, and it will only be necessary at this point to state the facts observed. In a considerable number of cases an attack of chorea comes on immediately after or during the course of an attack of acute rheumatism, or there is a history of rheumatic joint affection some time be-
fore. This does not however exhaust all the cases associated with rheumatism, for in some the joint affection is so slight that it passes unnoticed. Rheumatic arthritis in children is usually trifling compared with that in adults, and a slight painfulness in one joint with malaise and headache may be all there is to show for the onset of a rheumatic attack which yet ends in serious endocarditis. From these considerations it will be obvious that any statistics on the point will give numbers much below the actual but even so the percentage of cases showing a personal history of articular rheumatism is sufficiently high to be remarkable.

In a series of 80 cases Heringham obtained such history in 37 or 46.25 per cent; Garrod in a similar number in 36 or 44.76 per cent; Stephen Mackenzie in 44.76 per cent; Barlow in 60 per cent and Cheadle in about the same per cent. In my own cases there was a history of marked pains in joints and limbs or of definite rheumatic fever in 21 out of 42, while in many others frequent and persistent headaches were complained of, which in my experience are specially liable to occur in children of rheumatic constitution.

In a smaller proportion of cases there is a history of a preceding attack of scarlet fever. There were six cases with such history out of 42 in my series, but in only two did the chorea closely follow the scarlet fever, and even in these there was an interval of some weeks. In the others there was an interval of one to
It is significant that in four of these cases joint and limb pains were marked after the scarlet fever. James Priestley has recorded the cases of chorea which occurred after scarlet fever in the North Eastern Hospital London, in the years '94, '95, and '96. Out of 8,360 cases of scarlet fever there were only 13 in which chorea developed, which gives a proportion of 1 in 412, not certainly a large one; of these 13, five, or 39 per cent, had rheumatic manifestations, which is nine times the ordinary percentage of rheumatism in scarlet fever. Eight out of twelve of these cases had signs of heart disease. It is possible that the association of chorea and scarlet fever is an accidental one, and certainly observers are not agreed in considering chorea a sequela of scarlet fever. Henoch states that it is rare, he having only seen four cases in which the two diseases were associated. It would be impossible to decide the question without statistics of very large numbers, combined with a knowledge of the average incidence of scarlet fever and chorea in the child population. In all probability however, considering that rheumatism is also found associated with scarlet fever, these scarlet fever choreas should be regarded as rheumatic, and the figures just quoted, regarding the enormously greater percentage of rheumatism in scarlet fever chorea cases than in scarlet fever alone, bare this out.

In some cases chorea has been known to develop im-

mediately after a profound Mental emotion chief of
which is fright. The number of instances in which the
disease immediately succeeded this supposed cause is
not large, but a sufficient number has been recorded to
make it evident that the connection is more than a
mere coincidence. In other cases the disease develops
a short time after a fright or severe mental distress.
A history of fright was only obtained in four of my
cases and in only one did the chorea come on imme-
diately after; in a second there was an interval of two
weeks and in the other two the disease, already pre-
sent to a very slight extent, became much worse imme-
diately after. The proportion of cases in which mental
emotion, as an exciting cause, can be traced is various-
ly stated by different authors. Gowers puts it at be-
tween one fifth and one fourth, Osler 15.5 per cent,
Angel Money 25 per cent, Garrod 18.75 and Heringham
12.5 per cent.

Besides sudden and violent emotion we find in a
number of cases a history of overstrain of one sort or
another, such as worry over school lessons, too hard
work, or harsh treatment by guardians.

Imitation has often been said to be a direct cause
of chorea. The sooner this fallacy is removed from our
text-books the better. No doubt the nomenclature is
chiefly responsible for this mistake, for the dancing
mania originally called Chorea Sancti Viti largely
spread by imitation, as all hysterical affections do, but
that the malady we are now discussing ever arises from
this cause can certainly be denied. Almost all modern
writers agree in testifying to their never having seen a single case arise by imitation, and the fact that while large numbers of choreic patients are always under treatment in our hospitals it has never been known to spread to another inmate of the same ward, ought to be regarded as conclusive.

There is also no evidence to show that chorea is ever due to Reflex Irritation whether from intestinal worms or other causes. Such conditions are, of course, present in some cases, but the removal of the irritation, as the expulsion of worms, has not influenced the progress of the disease in any way.

Pathological Anatomy.

Our knowledge of the pathological anatomy of the nervous system in chorea may be stated in a very few words. No gross lesion has been found in either the brain or spinal cord, and in many cases microscopical examination fails to reveal any alteration whatever. Such small changes as have in some cases been found, may more properly be regarded as due to the abnormal overaction of the nerve elements, in consequence of the diseased condition, than as the original cause of the disease itself. When present, they consist for the most part in hyperaemia, minute extravasations and spots of softening, sometimes minute haemorrhages and embolisms, and periarterial exudations. These vascular changes have been found scattered through the brain and sometimes the spinal cord, being especially marked in the intracranial motor tract and its con-
nections. In rare cases where severe endocarditis has been present, considerable plugging of vessels has been found, but this can only be regarded as an accidental concomitant. In the nerve cells themselves, degeneration and vacuolation have occasionally been seen. These alterations being widely distributed throughout the brain and being by no means invariably present, cannot be regarded as affording any evidence as to the precise locality of the primary lesion.

Pathology. The cause of chorea must still be regarded as somewhat obscure, but I hope to show that there is sufficient evidence to justify the conclusion that the disease is the result of the blood state, which we are accustomed to call rheumatic, acting upon a previously prepared soil.

As post mortem examination affords us little or no help we are forced to look to the symptoms for indications of the diseased locality; and this, in spite of some arguments to the contrary, may unhesitatingly be placed in the brain. The frequent involvement of the mental functions, the fact that the disease may be entirely confined to one side of the body or beginning on one side subsequently extend to the other, the cessation of the movements during sleep, the frequent affectation of speech, the marked effect of emotional disturbance on the disorder, all point conclusively to an affection not of the spinal cord, but of the brain. The argument for localising the disease in the cord is largely based upon experiments on dogs suffering from "chorea," in which the disease was said to continue after division of the cord.
Canine chorea however is not the same disease as chorea in man. It is probable that the cord is, at least in some cases, secondarily affected and, though the variability of the state of the knee jerks makes it impossible to base any definite conclusion on an alteration in them, the fact that they are seldom found to be normal, argues the presence of some contributing affection in this situation. As to the exact seat of the disorder of function in the brain, though definite statements are impossible, it seems reasonable to suppose that this lies in the cerebral cortex. There is no evidence from the clinical signs that there is any distinct disturbance of afferent impulses and if I am right in my contention that interference with the co-ordinating mechanism plays no part in the production of the symptoms it is not necessary to look further back than the cortical cells, from which motor impulses proceed directly to the cord, for the primary lesion. It was at one time supposed that the central ganglia of the brain were the seat of the disorder, but more recent investigation has shown that they have not the close relation with the motor and sensory paths which was at that time believed; the Corpora Striata at least having no share in the machinery of co-ordination.

A theory was brought forward by Hughlings Jackson and supported by Tuckwell, Kirkes and others, that embolism of the parts in the region of the Corpus Striatum was the cause of chorea. In view of what I have already

1. cf. Foster's Physiology 5th. Ed. part 5.
said, it seems only necessary to point out, that it is impossible to look upon embolism of any part of the brain as a possible cause of chorea. First because, there is in the majority of cases no evidence of such process; and second because the cause of the supposed embolism, namely endocarditis, is frequently not present throughout the whole course of the disease and because when it is present it often develops after the onset of the chorea.

Of the real nature of the change in the cerebral cells we are entirely ignorant, but that it is not a very profound one is shown by the fact that in the vast majority of cases the condition is entirely recovered from, and that it does not produce any secondary nutritional changes in the muscles and nerves. In some way or other the normal resistance of the cells must be lowered, or there must be increased irritation, so that spontaneous, uncontrolled discharges of nervous energy take place. These discharges differ, apparently, from the spontaneous discharges occurring in epilepsy, in remaining limited to the area of the cortex corresponding at any moment to the muscular movement, that is they do not extend to contiguous cells as is believed to be the case in that disease. They are also intermittent and casual in character, and do not tend to recur in rapid succession. It must be remembered however that this change is not limited to the part of the cortex concerned with muscular action, but is present also in other parts notably that concerned with intellectual processes.
We have now to consider what is the cause of this alteration in function of the nerve cells, but in estimating the importance as a factor, of any external cause acting upon the cells, it is necessary to bear in mind the influence of predisposition on the part of the cells themselves, as an equally important factor.

Some authors, notably Sturges, have endeavoured to show that chorea is merely an exaggeration of the normal restlessness of childhood, the exaggeration being produced in debilitated and enfeebled children by emotional disturbance. Chorea, in fact, according to Sturges, is a purely functional disorder of the nervous system. In support of this he describes, in a very able manner, the gradual development of thorough control over muscular movement, pointing out that this control is naturally incomplete in childhood, and that choreic movements are exactly similar to the restless spontaneous movements, which are ordinarily seen in children. Furthermore, he maintains that children who develop chorea have generally suffered from harsh treatment or worry or have been overworked at school. Quote his own words: "Chorea is bred not born. The bulk of the patients are children who are alarmed or put upon by harsh parents, harsh lessons, school punishment, insufficient food or ill-usage of any kind." He considers also, that the fact, that of 216 cases, 155 were brought to hospital in autumn and winter and 80 in spring and summer, confirms his contention that unfavourable external influences are responsible for the development of the disease. As might be
expected he discounts the influence of rheumatism as a factor in causation and curiously enough, though admitting that of the rare cases occurring before the age of six "almost all are intimately connected with rheumatism," affirms that "in the majority of cases of chorea in this country, occurring at its common age say between six and twelve, rheumatism takes no part whatever." There is no doubt a considerable amount of truth in this theory, but that it embraces all the factors in causation, or is an adequate explanation of the origin of chorea, I would distinctly deny. In the first place, the facts stated by Sturges are not confirmed by other observers. The number of cases in which worry, or fright, or harsh treatment is found as an immediate antecedent, form only a small percentage of the whole, and other statistics do not show such a preponderance of cases in autumn and winter compared with spring and summer. I have furthermore been struck by the fact that the mothers of choreic patients brought to hospital are not commonly of the careless indifferent type, but are on the contrary usually more careful, more anxious than the average. Secondly, this theory affords no explanation of the cardiac complications of chorea, unless we suppose that a functional disturbance of cerebral action can of itself produce endocarditis.

We have seen that rather more than 50 per cent of all chorea cases suffer from endocarditis, while a smaller number develop pericarditis as well. In addi-

2. Sturges.
ion to this the post mortem records of fatal cases show endocarditis to have been present in 95.75 per cent, and as stated by Gowers, all patients having more than three attacks had signs of organic heart disease. This cardiac condition cannot therefore be accidental, and we are driven to the conclusion that, either chorea is a cause of endocarditis, or that there is a common cause underlying both conditions. That the latter is the true explanation we can hardly doubt, seeing that the endocarditis is sometimes anterior in time to the development of the chorea. (We have already seen that the endocarditis cannot be the cause of the chorea.) Furthermore our knowledge of endocarditis, and analogy from other diseases, leads us to conclude that no nervous influence or mechanical alteration can account for the development of an inflammatory affection of the endocardium, but that the cause is to be traced to some chemical alteration in the blood state; to the presence of some poison circulating in the blood. It is only reasonable therefore to suppose that such a chemical poison is the cause of both the endocarditis and the functional alteration in the nerve cells, which is the basis of chorea. Nor is it difficult to believe that a chemical irritant or poison could have this effect; the action of lead in producing wrist-drop, the paralysis following absorption of diphtheritic toxin, or the mydriasis due to atropin, these, and many others which could be cited, are examples of the profound effect exercised on parts of the nervous system by the introduction in-
to the blood stream of a chemical poison. Consideration
of such instances shows us also that the effect may be
produced and may remain a long time after the poison
has ceased to be absorbed. The injury or disturbance
of function once brought about is only very slowly and
gradually recovered from.

Having arrived at the conclusion that we must
regard the cause of chorea as in all probability a mor-
bid blood state we may now consider what the abnormal
element is. I maintain that we are justified, by the
facts, in the belief, that the morbid blood state is that
known as the Rheumatic. Gowers in his work on the dis-
eases of the nervous system argues that the common
cause of the endocarditis and the chorea is a blood
state allied to, but not identical with, that which that
which causes rheumatism. But I see no reason for the
supposition that it is a blood state differing from
the rheumatic. The manifestations of any disease are
never absolutely identical in two persons; we do not
know why in the case of two persons suffering from rheu-
matic fever one should have endocarditis and the other
escape, but we yet call the disease in both instances
by the same name. Nor can we say, why the rheumatic state
in one individual should produce tonsilitis and in an-
other pleurisy, nor why the incidence of rheumatism in
childhood should differ so much from that in adults;
and we may therefore surely argue that variations are
due, not to the cause in particular cases being differ-

1. The rheumatic form of multiple neuritis is
an example perhaps even more apt in this connect-
ion than those mentioned above.
ent, but to the different re-action of the organism.

It will be well to consider in this connection in what way the incidence of rheumatism differs in children from that in adults, and how far these variations are in favour of the connection of chorea with rheumatism. Those who deny this connection have too exclusively looked for manifestations of the rheumatic state in similar childhood, to those occurring at a later age. We owe to Cheadle the most lucid and admirable account of the differences at the two ages. Whereas in adults the most prominent signs are fever and an affection of the joints; in children the joint affection is usually evanescent and trifling, and may be absent altogether, while fever is proportionately less. The profuse sweating too, with the accompanying sudamina, are rarely if ever seen, and rheumatic hyper-pyrexia, generally so fatal in adults, is never known to occur in children. On the other hand, though what we are too much accustomed to regard as the cardinal symptoms of rheumatism may be absent or trifling, other serious manifestations are more frequently present. The endocardium is more frequently attacked in children than in adults, indeed cardiac inflammation is twice as common in children and while pericarditis, pleurisy and tonsilitis of rheumatic origin, are frequently seen at both ages, subcutaneous nodules, erythema exudativa and, I include, chorea are manifestations of rheumatism, very largely if not entirely, confined to

1. Coll. Invest. Record. vol. iv p. 71

The increased liability of the young heart to suffer from endocarditis is also borne out by Church's statistics of 700 cases of acute rheumatism. Below 30 the percentage of cases in which the heart was affected was 65.38 whereas between 30 and 50 it was only 25.72.
early life. These various phases of the rheumatic state may appear in varying order, they may occur simultaneously, in close sequence, or may be spread over a considerable number of years widely separated from one another, or any one of them may appear by itself alone. It is rare however that careful observation will not reveal the presence of more than one of these manifestations at some time or other but it can not be too much insisted upon that we may have to look to the subsequent history of the patient for other evidences of the rheumatic poison than the particular one at any moment exhibited. The instructive cases recorded by Cheadle show well these points. They show also that the endocarditis is not, as in adults, invariably an accompaniment of arthritis, but may occur some time after the arthritis, or may even precede it; and the same may be said of chorea. It is not denied that with the exception of tendinous nodules which are always rheumatic, the conditions mentioned above may and do arise independently of rheumatism, but rheumatism is their commonest cause. I consider that in the case of the endocarditis so frequently associated with chorea we are justified in assuming that it is generally rheumatic, that indeed it is by itself evidence of the rheumatic condition. It is universally admitted that rheumatism is the commonest cause of endocarditis and if we are able to exclude other known causes such as pyaemia and uraemia, we must either put down the unexplained cases as rheumatic, or invoke the agency of some totally unknown cause, which seems to be
both unscientific and unnecessary. We have seen alrea-
dy that chorea cannot be regarded as the cause of the
endocarditis and moreover in fatal cases the post-mortem
changes in the valves are found to be exactly simi-
lar to those occurring in endocarditis distinctly as-
associated with rheumatic arthritis. I hold therefore
that we must conclude that the association of endo-
carditis with chorea is evidence that in these cases
chorea is rheumatic. If moreover, chorea is really one
of the manifestations of the rheumatic state it is not
necessary in order to prove a rheumatic origin to find
evidence in every case of other manifestations. Joint
lesions of a certain kind are, even when standing by
themselves, allowed to be sufficient ground for forming
a diagnosis of rheumatism; similarly we must expect to
find a certain proportion of cases of chorea without
other history of personal rheumatism.

What however are the evidences for the close associa-
tion of rheumatism and chorea?

1. The family history of chorea cases shows evidence
of rheumatic fever in almost the same percentage of
instances as in the case of rheumatic fever itself.

2. In 45 to 60 per cent there is a personal history
of preceding rheumatic affection. It must be remem-
bered however that these figures must necessarily be
much below the mark. A. Because arthritis is usually
the only manifestation looked for. B. Because the arthri-
tis of childhood is often so slight that it is passed
unnoticed. C. Because chorea may come first and other
rheumatic manifestations later.

3. Rheumatic arthritis and chorea are frequently present at the same time, or the one may immediately succeed the other. — The cases of Bessie Q., Kate P., Eliz. S., Marian H., and Ethel L. in my series are examples of this, and innumerable instances are to be found in other records.

4. In over 50 per cent of chorea cases, inflammation of the endocardium leading to organic heart disease is present, and is almost invariably found in all cases having more than three attacks.

5. Chorea is found in association with scarlet fever, and rheumatism is the only other disease similarly associated; a large number of the post-scarlatinal chores being also rheumatic.

6. The sex incidence of rheumatism and chorea show a remarkable harmony. We have seen that chorea is immensely more frequent in girls than boys and it would seem from statistics that acute rheumatism is rather more common in males than females. — Combining three sets of statistics I find that out of 1489 cases there were 809 males and 677 females. Further examination of these statistics, however, reveals the fact that during the age at which chorea occurs the preponderance is the other way, there being more females affected than males, while during the period of eleven to fifteen, that is the most common age for the onset of chorea.

1. Cheadle — Church & St. Thomas's Hospital reports.
girls suffer from acute rheumatism nearly twice as frequently as boys; the numbers given in the Collective Investigation Record for this period being 47 girls to 25 boys.

7. The seasonal incidence of chorea and rheumatism have been shown to harmonize.

8. The geographical distribution of chorea and rheumatism very closely correspond.

These evidences of the close association of chorea and rheumatism are most simply explained by the supposition that both conditions are manifestations of the same morbid blood state, which owing to causes which we can only to some extent guess at, expresses itself now as an endocarditis, now as a chorea, and again as an acute joint affection. Nor does the existence of cases due apparently to the agency of profound emotional disturbance invalidate the argument for the rheumatic basis of chorea. The cases said to be due to fright show signs of endocarditis at least as frequently as others, and as instances of the close association with rheumatism, even in these, the two following records are of interest.

1. E.M. A case reported by Osler of nerve chorea ending fatally in a girl of 18 due apparently to fright, the disease having developed on the day immediately succeeding a fright. At an autopsy acute endocarditis was found to be present, with recent vegetations in the mitral valve.

2. The case of a boy J.T., reported by Cheadle. In Nov. 86, at the age of six he had an attack of chorea attributed to fright. In Aug. 87, he had a second attack
attributed again to a fright. In Nov. 87, he developed arthritis, and subcutaneous tendinous nodules, followed in December by endocarditis. He had a third attack of chorea in June 88, accompanied by a second eruption of rheumatic nodules, by arthritis, progressive endocarditis and pericarditis, which proved fatal in August of the same year.

Rather do these cases of fright chorea give us some indication of the causes which are at work in determining that the morbid blood state in any individual shall produce this effect upon the nervous system.

This leads us to the consideration of the second factor, which I have indicated as of equal importance with the morbid blood state, in the causation of chorea. That factor is an unstable condition of the nervous system. The common age for chorea is the age at which the greater part of the growth in bulk of the brain having taken place, the functions of control have not yet been fully developed. The normal restlessness of childhood is an evidence of this incomplete control in the motor regions, which in the region of the mental functions, expresses itself as excitability, want of persistence, and emotionalism. It is almost superfluous to remark that a function in an early stage of its development is more easily disturbed and perverted in its action, than when full maturity has been reached. In the marked preference of the disease for individuals of the female sex, we have further evidence of the action of this deficiency of nervous inhibition, for it will be readily ad-
mitted that women in general are much more liable than men to diseases resulting from diminution, or loss, of nerve control. There is thus a physiological basis for both the age and sex incidence of chorea.

Beyond however this instability natural to the age of childhood, we find in the great majority of choreic patients, to a greater or less degree, that special constitution which has come to be called the neurotic. We are but too familiar with the clinical picture which this term calls up. The eager, active intelligent, emotional child, whose self control has not kept pace with the development of his other faculties, is common enough at the present day and especially so in large towns. This explains the special distribution of chorea in these centres, for it is in such children that chorea is prone to development. In 84.2 per cent of my cases the children were described by the parents as specially nervous or excitable. Sometimes the motor functions are more markedly affected and the child is noted to have been always unusually restless and fidgety. More often the excitability is mental; the child's sleep is restless and disturbed; it is prone to night-terrors; is eager and quick at school; frets and worries over its lessons; is abnormally affected by any change in its surroundings; is easily alarmed and terrified. And here I may say that I consider that the influence of fright in the actual causation of chorea has been very much exaggerated. It has been regarded as an efficient cause by itself, has been so described in ever
text-book and fright choreas have been labelled as a separate class by themselves. That profound emotional disturbance and chorea are not unfrequently associated I do not deny, but that this disturbance can be the actual and sole cause I do not believe. The child with the neurotic temperament I have just described as one of the essential factors in the production of chorea, is the child who is liable to be frightened easily and to be for a length of time affected by a fright. What I consider the fright does in those cases in which the chorea follows as an immediate sequence, and these alone are the cases worth considering, is merely to precipitate an attack already impending, to temporarily lower the resistance of the cells so as to allow of the invasion of the disease. The frequency of this sequence has also it seems to me been overestimated. In only one of my cases was a history of immediately preceding fright obtained, even on enquiry. Previous worry and overstrain fall too into much the same category. Continued for some time they have the effect of lowering the tone, of producing that departure from the state of normal health which makes it possible for the morbid blood state to take effect. Some observers, laying much stress on this instability of the nervous system, have been content to look to the action of this single factor as sufficient explanation of the origin of the disease. In my view however it can only be regarded as a state of predisposition, a condition of preparedness in the soil for the implant-
To the influence then of these two factors, the rheumatic poison or virus circulating in the blood, acting upon a nervous system not yet fully developed and in a condition to be easily thrown out of its state of equilibrium, the onset of chorea is to be attributed. How much in any case is to be put down to one and how much to the other can hardly be said, but the relative importance of each must vary in different individuals. Just as in a case of septic infection the fate of the organism may be sealed either by the overwhelming amount or virulence of the poison or by the low resisting power of the organism, so here we find the evidence stronger now for one factor now for the other.

**Diagnosis.** The diagnosis of the disease usually presents no difficulty whatever. The characteristic movements, the age of the patient and the history of recent onset are usually quite sufficient for the recognition of the complaint in all ordinary cases. It is only in very slight attacks, or in those in which there is marked weakness with little or no spontaneous movement, that any difficulty is likely to arise. In the first, careful observation, particularly during voluntary action, such as holding the arms extended in front of the body while the eyes are tightly closed, will generally reveal the characteristic movements if the disease is present. In the second, the paralysis is rarely if ever complete; it is limited to one arm or one leg and does not involve the face, or the other limb on the same side;
and, if the child is carefully tested and observed, choreic movements will not fail to be detected in the weakened limb or elsewhere. With cases occurring in adults which are sometimes more difficult of recognition, we are not here concerned.

**Prognosis.** In the vast majority of cases the disease is entirely recovered from and the danger to life may be estimated from the fact that out of the 439 cases of the Collective Investigation Committee, there were only nine deaths. The prognosis in children is rather better than in adults, the chorea of pregnancy showing a mortality of from 20 to 25 per cent. In very severe cases death may sometimes result from the exhaustion consequent on the continued movements, combined with the want of sleep and hindrance to the administration of food resulting therefrom. The cardiac complications are rarely severe but occasionally, especially when pericarditis occurs, death may be due to this cause. Naturally this result is more common in recurrences than in first attacks. Generally speaking, if the child is obtaining sleep and a fair amount of nourishment and there are no urgent cardiac symptoms, no anxiety need be felt as to the issue. The mental disturbance and the muscular weakness as a rule pass away along with the disappearance of the movements, but when convulsions have occurred in the course of the disease they have sometimes been known to persist and pass into true epilepsy. In some instances the disease passes into a chronic state and may continue for years,
but these are rare and I have not met with any examples in my own experience.

Treatment. By far the most important indication for treatment is to place the patient as much as possible at rest. In even mild cases it is well to keep the child in bed for the first week, and in severe cases he should be absolutely confined to bed during the whole course of the disease. The exhaustion entailed by the constant movement demands that the patient’s strength should be husbanded as much as possible, and while at rest the movements are generally rather less. The effect of a few days rest in bed, when the disease is first recognised, is often most marked and continued, the movements remaining less violent after the child is allowed up. All causes of fatigue of any kind whatever, such as lessons etc., should be entirely prohibited, and anything which tends to irritate or excite, such as visits of friends, or exciting books, should be forbidden. In a word, the external conditions should be made as soothing as possible. The importance of obtaining a sufficient amount of sleep is very great and when this is deficient, or absent altogether, recourse should be had to the use of hypnotics.

The food should be simple and light, but nourishing, and, in cases with violent jactitation, must be given frequently in small amounts. If necessary such cases must be fed by tube through the nose. When the movements are very severe, measures must be taken to prevent the patient throwing himself out of bed, or injuring himself
against neighbouring objects. Attention to the skin is also required, as severe bed sores are apt to form.

The mere list of drugs which have been tried in the disease indicates by its length that no one of them is a specific. It is doubtful indeed whether the administration of any drug really shortens the duration of the disease. Its course is so variable that nothing but a comparison with a large number of cases treated by rest alone would afford decisive proof of such action in the case of any remedy, and evidence of this sort is not forthcoming. The mild cases of short duration will get well without any drugs whatever and the more persistent ones do not seem to be much affected by any medicinal treatment. On the other hand though we may not be able to cut short the disease, treatment appears certainly to have some influence on the severity of the symptoms. Of sedatives, Chloral, in my experience, certainly gives far the best results, and is especially indicated where there is sleeplessness. It may be given in doses of five to ten grains or more three times a day and is well borne by children. Under its use the patient becomes much quieter mentally, sleep is less disturbed and restless, and the movements are frequently less violent. The Bromides, contrary to what might have been expected, are of much less value and indeed seem practically to have no effect whatever. Opium also has been tried, but does not give such good results as chloral, while its disadvantages are greater. The Salicylates are not of any value whatever, as far as
the chorea itself is concerned. Considering its rheumatic basis this may perhaps be regarded as singular, but it is well known that the same absence of effect is observed in the case of endocarditis arising in the course of acute rheumatism. Their use is of course indicated in cases complicated with joint affection or rheumatic pains.

The drug which has been most extensively used and recommended, since it was first advocated by Romberg, is Arsenic. It certainly seems in many cases to do good, and under its use the symptoms frequently become less marked. That it is as valuable however as many believe, or that it should be employed as a routine in every case, seems to me distinctly doubtful. Nor do I see any justification for its employment in the large doses frequently recommended. Some authors advise that it be pushed till symptoms of intolerance are obtained and give ten, fifteen, or even twenty minims of the Liquor Arsenicalis three times daily. Apart from the fact that arsenical neuritis occasionally results from its use in this way, any slight advantage, which may be gained, is more than counterbalanced by the interference with nutrition and the debilitating effect produced by the gastric disturbance, which it sets up. If there were distinct evidence that the disease was actually cut short, it might be worth while, in spite of these consequences, but there is none to show that smaller doses do not have quite as much effect. It is even asserted by some that the administration of Arsenic actually
lengthens the duration of the disease and protracts convalescence, and I have been informed by one of the medical officers at Shadwell Hospital for Children, that he noted that cases under arsenic were longer in hospital, than those treated by rest in bed alone. Strychnine has been strongly recommended by Trousseau. I have certainly found it of value in the later stages of the disease and especially to aid in getting rid of the slight occasional twitches, which are apt to persist after the regular choreic movements have ceased. Further, its stimulating effect on the appetite and its tonic action on the heart are of real service. Alcohol is sometimes of the greatest value in severe cases, when the effects of the continual movements are telling on the strength. Given in considerable doses, it quiets the patient, improves the appetite, and generally aids nutrition. Of other therapeutic measures I have seen great good to apparently result from massage, in persistent severe cases. It should certainly be tried in those cases where other measures have failed, and the disease is serious and protracted. In grave cases too much importance cannot be attached to the necessity of giving as much nourishment as possible, compatible with the state of the stomach. The two last mentioned remedies alcohol and massage are probably of value in this way. Change of air and scene are certainly not to be recom
mended in the acuter stages of the complaint, but when
convalescence is fairly established it is of distinct
benefit. In these later stages, too, the patient's will
may be advantageously called into play, and he should
be encouraged to control the movements as much as
possible.
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Table Of Cases.
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<td>1</td>
<td>.P.A.</td>
<td>12</td>
<td>M</td>
<td>Very severe pain, Rhumatism, 6 months.</td>
<td>Growing pains last few months.</td>
<td>Nervous.</td>
<td>Very gradual</td>
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<td>.E.C.</td>
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<td>F</td>
<td>Suffer rheumatism.</td>
<td>- no right fright.</td>
<td>Very excitable.</td>
<td>In a few days before.</td>
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<td>Both equal</td>
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<td>Ischaemic mitral</td>
<td>Absent</td>
<td>About 6 weeks</td>
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**Remarks**
- Occasional twitching but no frank weakness noted. (June 1, 97)
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<td>Sudden</td>
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<td>46</td>
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<td>49</td>
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<td>51</td>
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<td>52</td>
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<td>Sudden</td>
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</tr>
<tr>
<td>53</td>
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<td>3/3</td>
<td>Sudden</td>
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<td>54</td>
<td>J. B.</td>
<td>6</td>
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<td></td>
<td></td>
<td>Sudden</td>
<td>Jan: 97</td>
</tr>
<tr>
<td>Side affected</td>
<td>Paralytic Symptoms</td>
<td>Cardiac Condition</td>
<td>Caudit. of Knee-jerks</td>
<td>Duration</td>
<td>Remarks</td>
<td></td>
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<tr>
<td>Both</td>
<td>-.</td>
<td>Systolic mitral.</td>
<td>Normal.</td>
<td>?</td>
<td>Slight movement present, weakness present.</td>
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<td>Both</td>
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<td>-</td>
<td>Exaggerated after 2 months.</td>
<td>2 months</td>
<td>Slight movement present, weakness present.</td>
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<td>Systolic mitral.</td>
<td>PAID at first.</td>
<td>3 months</td>
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<td>-.</td>
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<td>About both later.</td>
<td>2 months</td>
<td>Said case.</td>
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<td>Left mast.</td>
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<td>Slight attack.</td>
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<td>5½ months</td>
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<td>Right arm</td>
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<td>Inactive in left.</td>
<td>6 weeks</td>
<td>Council attending after 1st visit.</td>
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<td>-</td>
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<td>8 weeks</td>
<td>Council attending after 1st visit.</td>
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<td>About normal.</td>
<td>7 weeks</td>
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<td>Right arm</td>
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<td>Systolic mitral.</td>
<td>-</td>
<td>8 weeks</td>
<td>Council attending after 1st visit.</td>
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<td>Systolic mitral.</td>
<td>Exaggerated.</td>
<td>3 months</td>
<td>Council attending after 1st visit.</td>
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<td>Systolic mitral.</td>
<td>Normal, about during first month.</td>
<td>6 weeks</td>
<td>Council attending after 1st visit.</td>
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<tr>
<td>Both</td>
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<td>Systolic mitral.</td>
<td>Normal, about during first month.</td>
<td>6 weeks</td>
<td>Council attending after 1st visit.</td>
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<td>5 weeks</td>
<td>Council attending after 1st visit.</td>
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