C H O R E A.

A Study of 100 Cases.

Thesis for the degree of M. D.

by

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The material upon which this thesis is founded consists of 100 cases of Chorea that have been studied by me in hospital practice.

To the Physicians of the Bradford Children's Hospital and to Dr. Porter Parkinson, Physician to the Queen's Hospital for Children, London, I am particularly indebted for permission to make use of the cases.

Definition: Chorea is a spasmodic affection of the Nervous System characterised by involuntary movements, which are sudden, jerky and irregular, and by a variable degree of muscular weakness accompanied by incoordination of voluntary movements, and by psychical disturbances usually of the nature of habetude and mental weakness; to these may be added phenomena of arthritis and endocarditis. (Allbutt's System of Medicine.)

The name Chorea was originally used in connection with an epidemic of dancing mania, which seems to have been prevalent in the early part of the 15th century in Germany. An epidemic having occurred at Strasburg in the year 1418, the prefect of that city ordered/

ordered the sufferers to the Chapel of St. Vitus, that they might be healed by the saint.

The disease was from this time named "Chorea Sancti Viti." Subsequently many conditions in which spasmodic movements occurred were known under this name until Sydenham, in 1686, first described the disease I now propose to consider, and gave to it the name Chorea.

Etiology.

Sex: Of my 100 cases I found that :-

74 were Females.

26 " Males.

This practically gives the ratio of three Females to one Male. This agrees with most of the published statistics. For example Gowers found the ratio to be three females to one male out of 1365 cases. Osler gives 71 % Females and 29% Males. Holt says that it is more than twice as common in Females and Ashby and Wright give the ratio as five Females to two Males.

After puberty males are rarely affected and the ratio in favour of females increases annually.

Age: The youngest patient in this series was four years and nine months. From all accounts the disease seems to be very rare under the age of five.

Cases/

Cases of congenital chorea have been described but most observers agree that those were probably cases of spastic diplegia. Mackenzie in the Medical Annual for 1908 mentions a case in a child aged two years.

The average age in my series was 9.5 years and 94 per cent of the cases occurred between the ages of 5 and 15 years - the two limits being $4\frac{3}{4}$ years and 18 years.

Arranging the cases into periods of five years the following results were obtained :-

Years.	1-5	5-10	10 - 15	Over 15.
No. of Cases.	1	54	40	5

Osler out of 535 cases found :-

33 between the ages of 1 and 5
228 " " " " 5 " 10
212 " " " " 10 " 15
62 " " " " 15 " 20.

Ashby and Wright out of 252 cases found:-

Under 6 years	- 15 cases	- 3 boys	and 12 girls.
6 - 10 "	- 102 "	- 35 "	" 67 "
10 - 15 "	- 135 "	- 44 "	" 91 ".

Amongst the cases examined by the Collective Investigation Committee the results were :-

34 per cent/

34 per cent between 5 and 10 years of age.
43 " " " 10 " 15 " " "
16 " " " 16 " 20 " " ".

It will thus be seen that chorea is essentially a disease of childhood and early adolescence, the vast majority of the cases occurring between the ages of 5 and 15 years. According to Gowers not more than 5% of the cases occur after the age of 20, and then the disease is almost entirely confined to females.

Taylor however reports an apparently typical attack of chorea in a man aged 58, who was admitted to one of the London Hospitals, and rapidly got well under suitable treatment. This man had two attacks of acute rheumatism, and one other attack of chorea after the age of 50.

I have further divided my cases into males and females at the different ages. Thus :-

Age	Males	Females	Totals.
1 - 5	1	0	1
5 - 10	14	40	54
10 - 15	11	29	40
Over 15	0	5	5
Totals.	26	74	100.

Race/

RACE. According to Weir Mitchell the disease is very rare amongst negroes. For example the disease is said to be almost unknown in Cuba amongst the black children, whereas there it is quite common amongst the white children.

It is worthy of note in this connection that acute rheumatism seems to be especially prevalent in the West Indies.

HEREDITY. In only three of my cases could I get a history of chorea in one or other of the parents. It does not therefore seem to be a usually directly transmitted disease.

Three of the mothers of these patients had the disease. For example the mother of one had the disease at the age of 14 and had the second attack during the pregnancy of the patient. The chorea then went with the birth of the child.

Two other mothers had chorea in the pregnancy. None of the patients gave a history of the father having chorea, but one of them had an uncle with it. In 18% of the cases there was evidence of a neuropathic heredity.

The father of one patient had "always been nervous" while the mother was confined in an asylum, and two cases gave a history of epilepsy in the family/

family.

Of far more importance however is the question of rheumatism in the family, and that I shall take up later. Many families both have a rheumatic and a neuropathic heredity.

It has been said that the subjects of chorea do not appear to be subsequently prone to the occurrence of other nervous maladies.

More than one member of a family may be affected. Two of my cases had one brother and one sister who had suffered from the disease. Gowers mentions a case in which chorea occurred in three sisters.

Climate, Season, Locality.

Chorea is widely distributed all over the World and climate does not seem particularly to affect it. In my cases I was unable to trace any seasonal incidence but Lewis of Philadelphia has shown that the disease has a maximum incidence in March in America. In London, July and August is said to be the time for its greatest occurrence.

According to Lewis the highest spring point of the chorea tracing corresponds with cool weather and a low barometer and mean relative humidity tracing.

The greatest number of cases of acute rheumatism occur/

occur in the last quarter of the year.

Of my cases the admissions for the different months were:-

January.	8
February.	9
March.	6
April.	10
May.	6
June.	11
July.	8
August.	7
September.	11.
October.	9
November.	8
December.	7
	100.

Emotion and Allied Causes.

I found nine of the cases gave a definite fright as the reason for the attack. None of those cases gave any personal or family history of rheumatism, but all of them were delicate and certainly emotional children.

The chorea in most cases was not noticed for some days after the fright, but one case came on the day after the child had been punished at school.

Another/

Another case came on after a neurotic girl had been teased by her companions on beginning work in a large mill at Bradford. She had never had the disease before and had only been at work for three days. This turned out to be one of the most severe cases I have seen and was nearly fatal.

In the other cases the fright was not so obviously allied to the beginning of the disease, but they gave a definite history of fright within a short time of the beginning of the illness.

The question must therefore arise as to whether the fright can be looked upon as a cause, or merely one of the first symptoms of the disease. Many of those children were probably in a nervous fidgety condition long before the occurrence of the definite fright, and it was only due to the fright that the movements were brought out. Therefore the fright was probably rather the first symptom than the cause. Many children get frights, but few indeed get chorea. The fright can only act on a nervous system that is by some means lowered and weakened.

For example many of the cases of tubercular meningitis give a history of a blow or a fall to which the parents attribute the illness. But it is now known that the condition is the result of the tubercle/

tubercle bacillus, and that the injury only weakens the resistance of the patient and allows the tubercle bacillus to have the upper hand.

Grief, worry and various forms of excitement certainly are strong predisposing elements. It has been said to follow minor surgical operations and I have two cases that show this.

One patient developed chorea after the operation for tonsils and adenoids, and another after the extraction of a tooth.

Three bright and intelligent children gave overwork at school as the cause of their attack. I have noticed this fact that many of the children with chorea are smart and bright at school. The condition is said to be more frequent in the more highly cultivated races.

Reflex irritations are given by some as causes. If such irritations exist they should certainly be removed, but none of the cases in this series seemed to have such a cause.

Intestinal parasites are given as such causes, and several cases have been reported as stopping after the removal of tape or round worms.

Other reflex causes mentioned are phimosis, gastric/

gastric disturbances, adenoids, and dentition. All those conditions are very common in children from 5 - 15 years of age and frequently exist without chorea, so that I do not think they can be thought of as causes.

By some it is thought that imitation plays a part in certain cases. It is doubtful if this can be so, because the so called epidemic chorea is in most cases thought to be hysterical. Ashby and Wright state that they have noticed cases of chorea made worse by other cases in the same ward.

Errors of refraction are also given as a cause, but such conditions would be much more likely to produce some form of tic and not a true chorea.

One curious case has been reported after the use of iodoform. The chorea appeared after the use of iodoform for a fistula in ano, stopped when it was withdrawn and began again on its resumption.

PREGNANCY. I have seen cases of chorea in pregnancy, but this series does not contain a case. The relationship is important, because in many of those cases there is no other cause to be found. It seems to be determined by mental worry, overstrain, and shock. The determining cause can only be effectual/

effectual when it acts upon a brain weakened by the pregnant state.

Practically all the cases occur in women under 25 years of age. It usually comes on about the third month of the pregnancy, but it may occur at any time. It may follow abortion.

Wall and Andrews in 1903 published an analysis of forty cases of chorea in pregnancy.

It occurred in the first pregnancy in 18 cases. In 10 cases the first pregnancy was not attended by chorea, but the condition occurred in later pregnancies.

There was a history of previous chorea in 23 cases. Five of the thirty-seven women were unmarried.

There were five fatal cases among the forty. Two of those were spontaneous abortions, and in two fatal cases abortion was induced. One died from acute mania.

Month of pregnancy in which the movements began :-

Month.	1 - 2	2 - 3	3 - 4	4 - 5	5 - 6	6 - 7	7 - 8	8 - 9
Cases.	4	6	4	8	6	6	2	2
Per-centage	10.5%	15.8	10.5	21	15.8	15.8	5.2	5.2

From this table it will be seen that the movements were most noticed from the 4th to the 5th month, and that/

that after then the chorea was not so common.

In the majority of the cases the pregnancy continued undisturbed, and ended in a natural labour. There was only one case of mania. There is not necessarily chorea in the future pregnancies but Gowers mentions a case in which chorea occurred in five successive pregnancies.

The comparative frequency with which this condition occurs in the unmarried suggests that mental anxiety and depression play an important part as exciting causes.

RHEUMATISM.

This is far and away the most important etiological condition, much more so than all the others put together. Indeed many now believe that chorea is merely a cerebral manifestation of rheumatism and all, I think, are agreed that the connection is very close.

One of the great difficulties is in determining what exactly rheumatism is, where it begins and where it ends, and this doubtless accounts for the many different statistics published on this matter.

As long ago as 1802 Bright pointed out the close connection between chorea and rheumatism, and since that time much work has been done on this subject.

The/

The recent pathological investigations of Poynton and Paine, who have shown that an organism can be isolated from the joints in acute rheumatism, have rendered this almost certain.

They have cultivated this micro-coccus and have injected it into rabbits producing arthritis, endocarditis and chorea.

Amongst my cases I got a definite personal history of rheumatism in 39 cases. If I add to this conditions that are usually called rheumatic my percentage of cases was very largely increased.

I included in those rheumatic conditions tonsillitis, growing pains, and purpura of which there were three cases. I then got 30 other patients with a history of one or other of those conditions.

Adding those together I increased my cases of previous rheumatism to 69. This is, of course, a very large percentage.

To come to the Family History I found a history of rheumatism in one or other parent in 27 cases. Rheumatism is, of course, very common in this country, but to find so many with this definite family history seems worthy of note.

Of course the generation further back might have increased the number of previous cases, but it is/

is very difficult to go far back in the history of hospital patients.

On the other hand I have seen it stated as showing how common rheumatism is that if you take 100 apparently healthy people you will probably get a rheumatic family history in 25 of them.

35 of the cases had had former chorea.

In some of the cases signs of rheumatism came on under treatment. Three had pericarditis, several had endocartitis and a few tonsillitis.

One very important point had been brought out by Dr. Batten. From 115 cases of chorea he found previous rheumatism in 32%. Three years later he followed up those cases that gave no history of rheumatism.

He was unable to trace some of them, but he found that 11.3% had developed rheumatism. Three years later he again did the same thing, when he found that a further 9.7% had rheumatism.

From this it seems that the association of rheumatism is often subsequent to the chorea. The statistics published only give the number of cases with rheumatism previous to the chorea and therefore they must under estimate the proportion of choreic patients that have had rheumatism.

The/

The collective Investigation Committee of the British Medical Association found that rheumatism had preceded chorea in 26% of cases, while in 32 % the rheumatism accompanied the chorea, or occurred subsequently.

To those numbers 14 % must be added if patients who had been the subjects of vague rheumatic pains were included.

This makes a total of 72 % and if you add to this patients with a family history of rheumatism the number of cases becomes so high, that there seems no other explanation but that chorea , in the great majority of cases, is a manifestation of rheumatism.

In 1891 Sir Dyce Duckworth published a lecture on chorea in which he defined the condition as "a manifestation of the rheumatic habit on diathesis determined or precipitated oftentimes by some sudden emotion as fright, or occurring sometimes without such recognisable determinent."

It is not possible to produce evidence of rheumatism in every case. Many rheumatic ailments exist apart from joint troubles. Rheumatism is especially a disease of the motor structures affecting the heart and the joints. It is very apt to recur and /

and so is chorea.

Very varying statistics are given with regard to the number of cases that have had rheumatism, and they seem to vary as widely as 18% and 72%. It is obvious that the observers are not agreed on what evidence is sufficient for rheumatism.

Wood gives 18% of cases with previous rheumatism. Church 20% and up to at least 50% if the minor manifestations of rheumatism are taken into account.

Holt found out of 111 cases definite rheumatism in 63. In 41 the rheumatism occurred before the chorea, and in 13 the first evidence of rheumatism was coincident with the chorea, and in nine cases the rheumatism followed the chorea and usually within three months. He agrees that the evidence of rheumatism multiplies the longer the cases are followed up.

Lees gives rheumatism in at least 60% of cases, and considers that on bacteriological and clinical evidence chorea is cerebral rheumatism.

One of the most satisfactory proofs of the evidence of rheumatism is the presence of rheumatic nodules. Those I was only able to find in 4 of the cases.

But according to Thompson, nodules indicate that rheumatism/

rheumatism is not only present but active at the time, and they only last as a rule for a few weeks.

Taking all these points together it will be seen how close is the association with rheumatism. Fright, overwork, etc., are all powerful provocatives of the disease but in all probability they are not of themselves sufficient, in the absence of a rheumatic strain, to cause the condition.

Crocker showed that it was only in about 70 % of cases in which a definite tertiary syphilitic skin affection was seen that a history of syphilis or its early manifestations could be obtained.

The difficulty with rheumatism is at least equal to that with syphilis, so we may presume from analagy that many more cases of chorea have a rheumatic basis than even the figures show.

Morbid Anatomy and Pathology.

In this series of cases there were no deaths, and I have not seen a post-mortem examination after chorea. As it is so seldom a fatal disease the post-mortem changes found in the universally severe cases can hardly be supposed to represent the anatomy of the disease.

Endocarditis has been reported in practically all/

all the fatal cases, but no common cerebral lesions have been reported.

Hyperaemia of the brain and spinal cord with peri-arterial exudations especially in the deep portions of the motor tract have been noticed.

But in many cases no special microscopic or macroscopic changes have been found in the central nervous system.

Certain hyaline bodies, first described by Elischer, have been found in and around the arterioles and in the peri-vascular spaces. Those were termed by him chorea corpuscles, but they have been shown not to be peculiar to chorea.

There are many points in favour of the cerebral cortex being the part concerned in chorea, and especially the region of the Rolandic motor area. The abscence of sensory disturbances, the occasionally unilateral distribution and the exemption of the trophic functions of the lower motor NEURON all point to this.

The frequency with which attacks are excited by emotional disturbances, and the mental state during the attack, suggest that it is the highest part of the nervous system that is envolved.

Alteration in the appearance of the cells of the/

the motor cortex has been described, but other observers have denied this.

Perivascular infiltrations, capillary embolism, and small haemorrhages have been found. According to Taylor such lesions are the rule in subjects who have died of any infective disease, and this fact also helps to bring chorea and rheumatism together.

Changes in the heart have been found in practically all the fatal cases. In 1891 Sir Dyce Duckworth reported a fatal case, where after death no changes were found except in the heart. Marked endocarditis of the mitral valve was present. There was no sign of cerebral embolism.

Goodhart and Still report 28 fatal cases in thirty years from the statistics at Guy's Hospital; 25 of the deaths were in females. Endocarditis of mitral valve was present in 26 of those cases. It was doubtful in one, and absent in the other.

In those cases the mitral valve was affected alone fifteen times; the aorctic and mitral valves together nine times; the aorctic valves alone four times. Pericarditis occurred with the endocarditis on six occasions.

Carpenter (Lancet Nov. 1907) reports a fatal case/

case of Chorea with double optic neuritis. Thickening of the mitral valve, and a brain normal to the naked eye were found P.M.

There are three chief theories as to the cause of chorea:-

1. The embolic theory.
2. That it is a functional brain disorder.
3. That it is an infectious condition and due to a special organism - the diplococcus rheumaticus.

I. THE EMBOLIC THEORY. This is now more or less abandoned, for it will not account for many of the cases. It depends for its existence on the presence of endocarditis, but this condition cannot always precede chorea. The condition was supposed to be due to cerebral emboli, that had escaped from vegetations on the cardiac valves.

This theory received support from the experiments of Rosenthal, who artificially produced choreiform movements in animals by injecting fine particles into the carotids.

A few fatal cases have been found with this lesion. Duckworth reports an interesting case in this connection where hemichorea came on during an attack/

attack of rheumatic fever with mitral disease, and was afterwards recovered from. He believed that the condition was due to embolism.

Duckworth also points out that, where those emboli have been found after death, it could not be known whether the plugs were emboli or arterial thrombi.

It would be remarkable he says, if this tendency to embolism was confined to the cerebral vessels alone, and such common sites for embolism as the spleen and the kidneys should remain free - and this is the case.

Again in endocarditis with embolism chorea is not met with.

Simon reported a case in 1907 that favoured embolism, where a girl aged 22 became hemiplegic and aphasic. Both conditions were present five months later.

THE NERVOUS THEORY. Until the discovery of the diplococcus rheumaticus this was the theory that seemed to account for most of the cases. This theory is largely based on the appearance of the disease during the years of active growth, and on the complexity of nervous disorders which seem to embrace the condition.

Choreics - according to Church - occasionally show some of the/

some of the stigmata of degeneration. The tendency to a hemiplegic distribution seems to point to the condition being more likely due to some cerebral condition than to an organismal one.

Is the lesion an irritative one or a paralytic one? From the fact that the movements are so chronic it seems unlikely that the lesion is irritative.

On the other hand a paralytic lesion affecting the highest and most recently developed centres gives an easier explanation of some of the phenomena. The paralytic theory explains better the long duration of the movements, with their tendency to recovery, and also the fact that at times they become habitual and may be improved by re-education.

Osler seems to consider that chorea is a functional brain disorder affecting the nerve centres controlling the motor area. Of the nature of this derangement nothing is known, nor whether the changes are primary and the result of a faulty action of the cortical cells or whether the impulses are secondarily distributed in their course down the motor path.

The predominance of the disease in Females, its onset at a time when the brain is developing, are facts which Sturges has urged in favour of the view that chorea/

chorea is an expression of functional instability of the nerve centres.

ORGANISMAL THEORY. This theory depends for its origin on the discovery of the diplococcus rheumaticus in the brain of fatal cases of chorea.

In 1891 Pianese, an Italian, cultivated a bacillus from the brain and spinal cord of a fatal case of chorea. He cultivated this and injected it into animals producing apathy, tremor, convulsive movements and finally death. He then found the organism entirely confined to the animals' nervous system. As far as I can find no one has confirmed this.

Other organisms, especially staphylococci, have been isolated by other obeervers with no definite results up to about 1899. At that time Wasserman produced with constancy a polyarthritis in rabbits by inoculation of a diplococcus that he isolated from the cerebro-spinal fluid and inflammatory exudations of a patient, who had died from acute rheumatism and chorea.

In 1900 Poynton and Paine confirmed this, and further demonstrated a diplococcus in the pia mater and brain of a fatal case of chorea. In that year they also found that inco-ordinate movements resulted from the/

from the inoculation of this diplococcus into a rabbit.

These experiments were confirmed by Beattie who isolated this diplococcus from the knee-joint in a case of acute rheumatism. He cultivated it and injected it into a rabbit, and produced arthritis and choreiform movements.

In 1905 Paine and Poynton published a further paper with three fatal cases giving further confirmation of the above statements.

The first fatal case was in a girl, aged seven. Death occurred on 17th day of the illness, and she had had acute rheumatism. After death, pericarditis, endocarditis and myocarditis, were all well marked. A pure growth of the diplococcus was obtained from the heart's blood, its presence was demonstrated on the pericardium, in the pia mater, and in the brain. It was cultivated and on injection into a rabbit it produced arthritis, endocarditis and chorea.

Second fatal case was a boy, aet 14, who had been ill with acute rheumatism for two months. He was admitted to hospital with pericarditis. He gradually got worse and died. The interesting point in the case was that after admission to hospital he developed violent chorea. The bacteriology results were similar to the first fatal case.

In/

In those rabbits the diplococci were found in pia mater, and in the endothelial cells of the blood capillaries.

One animal showed endocarditis, arthritis and chorea. Another had the organisms isolated from the cerebro-spinal fluid, but it showed no sign of chorea although arthritis and endocarditis were present. A third rabbit showed weakness of the hind limbs, which it was thought might be due to chorea paralytica.

Poynton and Paine admit that they cannot prove all cases of chorea to be rheumatic, for they consider that other affections of the strepto-coccal type may produce the condition.

When no history of rheumatism can be got they consider that chorea is then the first manifestation of rheumatism. They consider that fright, pregnancy and those other conditions can only act on a brain that is weakened by the rheumatic diathesis.

To sum up Poynton and Paine have shown :-

1. They have isolated and cultivated the diplococcus from the cerebro-spinal fluid in cases of fatal rheumatism in some of which chorea was present at the time of death.
2. Twitching movements, arthritis and endocarditis, have/

have been produced by the intra-venous injection of this organism.

3. They have found this diplococcus in the pia mater and brain of fatal cases of chorea.

Vaccination has been tried in chorea by Fordyce. He obtained the organism from the knee-joint in a case of acute rheumatism. He used his own serum as a control.

In eleven cases of chorea he tried the index against this organism. In ten cases when the test was done soon after the patients' admission to hospital the index was low, and when it was done later, with the patient in a better state of health, the index was high. These facts support the theory that chorea is due to the diplococcus rheumaticus.

Other points suggesting the infective origin:-

I. Endocarditis is admittedly an infective condition. It has been shown that this is present in practically all the fatal cases, and it is probably present during life more often than can possibly be recognised. At all events the endocarditis found in cases of chorea is indistinguishable from the endocarditis of rheumatism both clinically and morphologically.

A case of chorea has been reported with streptococci/

streptococci present in the blood, which failed to yield to any treatment until anti-streptococcic serum was given and after fifteen injections of this the chorea stopped.

2. The age at which chorea occurs is similar to the susual age for infectious diseases. Chorea is most common from about 5 - 15 years of age and it is just at this time that patients are most liable to infectious disease.

3. The clinical course in a case of chorea resembles the course of an infectious disease somewhat. It may also be associated with infectious disease.

It is possible, says Lees, that various microbes and toxines, and perhaps sudden emotional disturbances may affect the nutrition of the cortical cells in a way similar to the altered nutrition caused by the rheumatic toxine.

If the condition is due to the diplococcus rheumaticus it is still unknown whether this organism acts locally on the cortical cells, or by means of a toxine. Probably both conditions occur.

SYMPTOMS.

The onset of symptoms is as a rule gradual, but especially if fright has had anything to do with the case, it may be comparatively sudden.

In/

In my cases the onset was very varied as regards time. For example in one case the movements were said to have been noticed off and on for two years, and in another the condition was noticed the day after the extraction of a tooth. There were all sorts of times between these two extremes; some of them had attended the out-patient department for two or three weeks and were only admitted to hospital on account of showing no improvement or getting worse. As a rule the cases with the sudden onset were the severe ones.

When the onset is gradual the appearahce of the movements may be preceded by mental and psychical disturbances in the patient. For example they become more nervous, more irritable, and often laugh or cry without any very apparent cause.

They become more clumsy in their movements and actions, perhaps let fall objects they are carrying or overturn glasses at table. The frequeht reprimands for such faults often increase the condition..

From day to day the movements become more noticed and the patient is then brought for advice. The movements begin as a rule in the face, and then they spread/

spread to the limbs and trunk. There are four chief classes of symptoms :-

1. In voluntary spontaneous movements.
2. Weakness of voluntary movements.
3. Loss of precision in voluntary movements.
4. Emotional and Psychical changes.

I. Involuntary Movements.

These are always irregular as regards time and as regards the nature of the movement. The movements begin suddenly and end suddenly, and each movement seems to differ from the one before. Most of the movements are complicated movements envolving several muscles and usually more than one joint.

At certain stages these movements resemble very much certain voluntary movements, for the child is often thought to be fidgeting and may be punished for it, when she absolutely cannot help it.

This similarity, it has been suggested, must point to the seat of the disease being in the neighbourhood of the Rolandic Motor area. In the face the movements are never confined to one side, even in hemichorea, according to Gowers. In all the severe cases there is difficulty with swallowing and with speech.

The way the tongue is protruded is often characteristic/

characteristic. It is shot out very suddenly, kept there some time, and then just as suddenly withdrawn, often just in time to escape being caught by the closing jaws.

In the upper extremities the movements begin usually in the hands, and often in the thumb. The thumb may begin to fidget about, then the fingers, wrist, forearm, etc. All sorts and all possible movements of the forearm and the upper arm may occur.

The clumsiness of the choreic patient is thus described by Sydenham; "If a cup of drink be put into his hand, he represents a thousand gestures, like jugglers, before he brings it to his mouth; for whereas he cannot carry it to his mouth in a right line, his hand being drawn hither and thither by the convulsions, he turns it often about for some time, till at length happily reaching his lip, he flings it suddenly into his mouth and drinks it greedily as if the poor wretch designed only to make sport."

The lower extremities are, as a rule, less severely affected than the upper. The gait is often clumsy, and in bad cases walking is not possible.

Respiration often becomes jerky and irregular. The movements are spasmodic, and I have often noticed loud/

loud sighing at the bedside of a choreic patient. The breath seems to be taken in rapidly, held for some time, and then let go with a loud sigh.

The trunk muscles are always envolved to some extent, and writhing, wriggling movements are very characteristic.

Sometimes the movements are confined to one side, the so-called hemichorea. There were very few of my caees that showed this markedly, but several began as hemichorea, and in fourteen of them the movements were always more marked on one side than the other.

The involvement of the face and trunk is always bi- latteral according to Gowers. In addition there may be marked loss of power in the affected parts.

The movements may be so violent that unless precautions are taken the patient will seriously damage himself. All the bony prominences must be carefully wrapped up in wool and bandages, and the bed padded in such a way that as little damage as possible will be done to the patient. The movements of the trunk may be so violent as to throw the patient out of bed.

The movements always cease during sleep and frequently/

frequently the patient is much quieter for some time after a sound sleep. Mental excitement and any attempt to perform a voluntary movement always excites the movements.

2. Weakness of Voluntary Movements.

In slight cases there may be no obvious change in this direction, but in certain cases this is very well marked. The weakness begins to be noticed a short time after the beginning of the illness.

It may be observed in one limb or on one side of the body. I recently saw a child in the out-patient room, where the choreic movements had got much less in the right arm than in the rest of the body, but there was marked paresis in this arm and the arm was hanging loosely by the side.

According to Risien Russell the muscular weakness may be very profound, e.g. in chorea paralytic, which is commonly hemiplegic.

There is a definite paresis, but never complete loss of power. This may come on gradually during an attack of chorea, or it may be seen before the movements have been detected. The paresis may persist for a long time, but it is said to be always recovered from. Osler mentions a case where wrist drop/

drop persisted for two years.

3. Loss of precision in Voluntary Movements.

This may be one of the first sysemptoms seen or it may be one of the last to remain. It is best seen in the movements of the hand and forearm, which lack their usual precision.

There is marked delay in performing certain acts. The motor centres do not seem to respond properly to the dictates of the will. These acts are responsible for the dropping of articles that are being carried, and for the failure to pick up with ease such things as pins on the floor.

4. Emotional and Psychical Changes.

These are usually very slight, but Osler remarks that it is always a wise plan to tell the parents of the patient that it is not the muscles alone that are affected, but that the general irritability and change of disposition are part of the disease.

The child is apt to become in many ways troublesome, the result of the toxine that is causing the disease.

Delirium may occur in very severe cases. Fortunately this is rare but it is always of grave omen. Mania has also been described in the adolescent and adult cases. Here again the prognosis is grave/

grave, but if the chorea can be checked the prognosis, as regards the mental state, seems to be good.

Sensory disturbances are rare. Pain in the limbs is seldom complained of, but headache may be a troublesome symptom.

Gowers holds that if there is any impairment of sensibility it is probably a hysterical condition. In the great majority of cases there is no affection of micturition and defaecation, but I have seen one very severe case of chorea paralytica where the motions were passed involuntarily. Abrasions may be seen in very severe cases.

Reflexes. Skin and deep reflexes are as a rule normal. I found all sorts of conditions of the knee jerk. In some cases it was apparently absent, and in other increased. Garrod states that diminution of the knee jerk is the common condition. In a few cases the knee jerk seemed unusually prolonged. It has been stated that arsenic might abolish the knee jerk, but it is doubtful if this could occur without other signs of arsenical poisoning.

Circulatory System. Symptoms of derangement here are quite common. In my cases I found :-

(1) Fourteen cases with cardiac dilatation.

(2) Eighteen cases where there were cardiac MURMURS propagated /

propagated in various directions.

(3) Twenty-two cases where systolic murmurs were present without propagation or any dilatation of the heart.

How many of those twenty-two cases with the non-propagated systolic murmurs were haemic it must be very difficult to say. In twelve of them the murmur was loudest over the base and probably was haemic.

One would need to have the patients under observation during convalescence for some time to really be sure whether a murmur in the course of chorea was organic or not. Endocarditis may occur without a bruit.

In the other cases there were some slight changes in the regularity of the heart beat, and in most of them the systolic murmur was loudest over the apex.

There were five cases of mitral stenosis, and one case of aorctic regurgitation in a boy. This latter was the only case in which the aorctic valve was affected.

Many of these mitral murmurs were harsh and were propagated into the axilla, and the most common murmur/

murmur was over the mitral valve. According to Gowers murmurs at the aorctic valve are very rare and he only found such murmurs present twice in 250 cases.

Endocarditis of chorea is invariably of the simple form, and in itself is not dangerous. But as can be seen from those cases it leads to those sclerotic changes in the mitral valve, which lead to incompetency etc.

Osler quotes important statistics bearing on this :- He examined 140 patients more than two years after recovery from chorea and he found :-

51 had a normal heart.

17 had functional disturbance.

72 had organic disease.

From this it will be seen how common and how serious are the cardiac complications. Practically all the deaths are due to this.

Some degree of myocarditis and dilatation, says Taylor, are almost invariably present when endocarditis exists. It must be remembered that endocarditis may give very indefinite signs of its existence, and many cases of chorea have been reported as giving no evidence of a murmur during life, and

yet/

yet after death the autopsy revealed endocarditis.

There is seldom cardiac pain or palpitation complained of, but three cases in this series had pericarditis.

The onset of pericarditis may be very insidious, but sometimes the signs are characteristic. It is usually accompanied by some degree of endocarditis and myocarditis. In the first place there is a rise in temperature and then a to and fro shuffling sound may be heard over the base of the heart, and later the characteristic friction sound comes on.

The pulse rate may be increased and there may be much praecordial pain with shallow short breathing and an anxious look.

Fluid may be effused into the pericardium, and then the heart dulness is increased on percussion. As the fluid increases the cardiac impulse moves upwards and outwards, but the friction sound does not necessarily stop with the onset of the fluid. In the three cases of this series one had pericarditis with effusion. All gave a rheumatic history, and all were recovered from.

Myocarditis also occurs as part of the toxaemia and it is doubtless the result of this that the dilatation of the heart occurs. The inflammation

which attacks the pericardium probably at the same time attacks the deeper structures.

The symptoms are very insidious, but it probably causes more feebleness of the heart's action and a small irregular pulse, and it will increase the dyspnoea. But its share in all this when pericarditis and endocarditis are present will be very difficult to determine.

PULSE.

Perhaps the most common cardiac change was irregularity and rapidity of the heart. Omitting those cases that showed signs of other cardiac trouble this was present in fourteen cases.

This irregularity might be functional and it has been attributed both to chorea of the heart muscle, and also to the results of the irregularity of the respiratory movements upon the heart.

HAEMOPOIETIC SYSTEM.

There is no special change that I could make out here, except in some cases there were signs of simple anaemia.

T. R. Brown (Internat. Clinics. Vol XIV) reports an eosinophilia in chorea of from 5.9%, which he says is constantly present.

CUTANEUS. AFFECTIONS.

These support the connection with rheumatism. Erythema nodosum, purpura, of which I had three cases, and subcutaneous nodules are all seen at times. Pigmentation of the skin is also sometimes seen, but it is said to be generally due to the arsenic given in the treatment.

URINARY SYSTEM

A pigment called uro-haematoporphyrin has been discovered in the urine of patients suffering from acute rheumatism. This was found in 14 out of 20 patients suffering from chorea by Garrod.
Rotch has stated that albumen and blood are frequently present in cases of chorea that have been treated with arsenic, and he looks upon the presence of albumen as one of the first signs of poisoning by arsenic.

Attention to this point has been paid during the last year in the Queen's Hospital, London, but in none of the cases was albumen or blood found.

TEMPERATURE - In uncomplicated cases, and in all the mild cases there is usually no fever. In the severe cases and in maniacal cases the temperature is always raised. Rheumatic complications are the common reason for the raising of the temperature, and/

and when this occurs it is always an anxious symptom. It may be the beginning of a tonsillitis, or more likely an endocarditis or a pericarditis.

DIAGNOSIS.

Ordinary cases do not as a rule present much difficulty in the diagnosis, for the nature of the movements at once suggests the disease, but where the usual symptoms are masked by paresis or mania the difficulty may be very great. Gowers has remarked that it should always be borne in mind when a child between the ages of seven and twelve years is said to have gradually lost the use of one arm, the disease is most often chorea.

Maniacal chorea might easily be mistaken for acute mania, but the history of the case will usually reveal that the movements of chorea were present before the mania, and also even during the mania certain characteristic movements may be seen.

Hysteria, as is well known, may simulate any disease, and diagnosis between this and chorea may be extremely difficult. According to Risien Russell the movements in hysteria are of a much more rythmical and monotonous character. But that genuine chorea may be seen in hysterical subjects is not surprising when/

when the close connection that both diseases have with nervous heredity and emotional influences is remembered.

Chorea paralytica may be very difficult to recognise especially when the child is seen for the first time with a paralysed arm or leg.

In this connection an interesting case has been reported by Dr. Porter Parkinson in 1904. He had a boy, aet 10, admitted to hospital with paresis of the right side. The history was indefinite, but he was said to have been ill for two weeks.

His heart was not enlarged, but there was an apical conducted systolic murmur. Reflexes were normal, and there were no electrical changes. After about a fortnight well marked chorea began, and in three months time he left the hospital quite well.

This case had been diagnosed as cerebral embolism, but the absence of any alteration of the reflexes suggested that this was not the cause.

I have recently seen another case of very marked chorea paralytica. This patient was admitted to hospital three weeks after a former attack of chorea. The chorea was then very well marked and there was difficulty in feeding. This was the third attack of/

chorea. There was no history of rheumatism, but the heart was affected and the pulse was about 112. The patient remained in much the same condition for about five weeks, when the movements got much less, but other alarming symptoms came on.

The mind became affected and the patient appeared to hear and understand nothing. Her whole body was paralysed and she was unable to move a hand or foot. The abdominal movements of respiration were reversed, showing paralysis of the diaphragm. She had to be fed by the nasal tube and she passed all her motions under her.

For ten days the patient did not speak, but at the time of writing the condition is slowly improving and she now takes notice of things going on around her. The paresis is not so marked and there is now bladder and rectal control. At no time did this patient have any arsenic.

It is those cases of chorea paralytica that are likely to give the trouble in diagnosis, but other diseases that produce movements and tremors may be difficult to distinguish.

Athetotic movements are continuous, uniform, and as a rule bi-latteral. They are usually confined to the fingers and toes. The movements of chorea are/

are much quicker and the contractions of the muscles briefer.

Friedreich's Ataxia might be difficult to distinguish, but here the family history of the condition, the gait, the speech and the presence of mystagmus should differentiate.

Various habit spasms must be distinguished, and there is a condition very like chorea known after poisoning by hyoscine. Various choreic movements are seen in certain cases of organic disease. This occurs in cerebral di-plegia, but as this condition is congenital, the movements occur from birth.

PROGNOSIS.

In the great majority of cases this is quite good. All the cases in this series recovered, and even paresis in chorea is, as a rule, quite well recovered from. Dangers are :-

1. Exhaustion from want of sleep.
2. The never ceasing violent movements.

The danger is very great when those movements cannot be controlled, but there many means of treatment at our disposal, and it is but seldom that some of them do not act.

The milder cases as a rule get quite well after isolation/

isolation, rest and good feeding. In pregnancy the condition is serious, and I have already given a list of 40 cases with five deaths.

The British Medical Association Investigation Committee found the death rate to be 2 % out of 437 cases. The condition is said to be more serious after puberty, but the statistics from the hospital for Nervous Diseases in London, between the years 1888 and 1903 do not bear this out.

These statistics after puberty presented no peculiarity either as regards severity or duration of the disease.

The more severe the attack the longer will be its probable duration. Recurrent attacks seem to be less severe than the first attacks. Gowers says death in a recurrent attack is very rare.

Endocarditis does not seem to affect the immediate prognosis, for embolism is very rare, but it may lead of course, to very serious cardiac changes.

Hyperpyrexia is very rare in spite of rheumatism and chorea being so common together.

COURSE and DURATION.

The disease tends to a spontaneous cure, but the duration of it is very variable. I found that the average/

average stay in hospital of these patients was 45.8 days, or a little more than six weeks. The following table shows this :-

Age.	1 - 5	5 - 10	10 - 15	Over 15
No. of Cases	1	54	40	5
Days in Hosp.	35.	45.5	54.2	55.

This shows that the older they were the longer they remained in hospital. The two extremes of duration in hospital were two weeks and about five months. The child that was in for two weeks was simply a mild, uncomplicated, recurrent attack. The patient that was in hospital for five months was a girl of sixteen who for some weeks was very ill with endocarditis and pericarditis.

Gowers says cases that last for more than a year are not rare, and slight cases with many remissions may last for many years.

The legs are as a rule the first to recover, then the arms, and lastly the face. If you grasp the hand of a convalescent patient you often feel slight tuggings remaining, especially if the child is agitated.

RECURRENCE.

This is quite common. Out of my cases thirty-five had an attack before. This is approximately onethird/

one-third of them.

One of the cases was having the sixth attack in nine years'time. Of the recurrent cases 26 were Females and nine Males :-

15 Neither gave a history of rheumatism nor had any heart trouble.

8. Both gave a rheumatic history and had heart trouble.

7. Gave a rheumatic history but had no heart trouble.

5. Had heart trouble without a definite rheumatic history.

From this it will be seen that the recurrent attacks did not bear any more relation to rheumatism than the first attack.

Three of the cases had "an annual attack" in the summer, but they were not very serious. Recurrence within a few weeks of an attack should be regarded as a relapse. A second attack seems to commonly come on in a year.

In many cases no particular cause for the recurrence was ascertained, but the patient was probably in a weak state.

Heart troubles may occur just as in first attacks, but if there has been no cardiac lesion before the second attack, the heart will probably remain free.

Osler found among 410 cases that :-

Two attacks had occurred in				110.
Three	"	"	"	35.
Four	"	"	"	10.
Six	"	"	"	3.

TREATMENT.

The first important matter to procure in all forms of the disease is physical and mental rest, and freedom from all sources of trouble and worry. Every exciting cause must be sought for and combated if possible. Even in the mildest of cases it is probably wisest to begin the treatment with at least a few days rest in bed, provided this can be carried out without increasing the fretfullness of the patient.

If the patient is a child she must be removed from school and lessonsstopped. She must not be scolded for her faults, or subjected to the possible teasings of other children. Her surroundings ought to be as bright and light and happy as possible.

In mild cases if treatment of that kind can be adopted together with plenty good food and some such simple tonic as Syr. Ferri Phos. Co., the patients would soon as a rule be well.

Such conditions are practically impossible to carry/

carry out amongst hospital out-patients and therefore the majority of such cases get worse, and finally have to be admitted to hospital.

Improvement in the condition of the bodily nutrition is very important. Most of those hospital children are ill-fed and the effect of good food on the course of a mild attack is often very marked. It should be impressed on the parents of the child that the ordinary diet should be supplemented with as much milk and farinaceous food as possible.

All causes of ill-health have to be sought for and remedied. Such conditions as dyspepsia, constipation, phimosis, and possible intestinal parasites should be attended to.

In the cases where the movements are more severe the patient must be admitted to a hospital. Care must be taken that the patient is in no way injured from the violence of the movements. If necessary the child must be put on a water bed, and the sides of the bed protected by pillows etc. The bony prominences will also need to be wrapped up in cotton wool and bandages.

If the case is so severe as this the patient will probably be unable to feed herself. She must then be fed by a spoon, or if swallowing is impossible, by /

by means of a tube. This must be done regularly so that the patient gets plenty of nourishment. It is very important to carry this out because one great cause of sleeplessness is want of food.

In the severe cases china or glass feeding vessels should not be used on account of the danger of their being broken by closure of the patient's jaws. Some form of enamel cup is safe.

When the child gets to hospital she must be put to bed in as quiet a part of the ward as possible. If the movements are severe the patient would be better isolated either in a room by herself, or, if that be not suitable, she can be isolated by means of screens at the end of a ward.

Isolation is a valuable remedy and I think I have seen as much good from this together with good feeding as from most of the drugs. Warm baths, if they do not frighten the child, are helpful and soothing.

DRUGS.

There is no specific remedy for the treatment of chorea, but there are drugs that can modify the severity of the symptoms.

There are two great classes of drugs used :-

1 Tonics.

2 Sedatives.

On account of the intimate relationship with rheumatism the salicylates and aspirin have been very largely used, but they do not seem to have any special effect on an uncomplicated case.

The salicylates do not seem to have more effect on an ordinary case of chorea than they have on a cardiac condition that is due to rheumatism, except in the complications of endocarditis and pericarditis where they are of value.

It is worthy of note that Lees says that sodium salicylate must be given in large doses. His method is as follows. To a child from 6 - 10 years of age he begins by giving ten grs. every two or three hours. He always gives with this double the dose of Soda Bicarb. The doses are gradually increased till signs of poisoning arise. He has given a patient as much as 200 grs. in a day with good results. One of the first signs of poisoning is air-hunger.

Lees argues that as in syphilis of the brain large doses of anti-syphilitic remedies are needed, so in rheumatic affections of the brain large doses of anti-rheumatic remedies should be given.

Arsenic. This drug has long been used and with benefit/

benefit in many cases. It was used in thirty-seven of these cases, and the method of its use was as follows.

The patient began with minims two of Fowler's solution three times a day well diluted after food. The dilution of the arsenic was minim one to one dram of water, and it was always given in the same dilution at whatever dose. The dose was increased by one minim every other day till the patient was taking ten or twelve minims three times a day. It was always stopped on the appearance of any sign of poisoning, the usual signs of which are itching of the eyelids, flushing of the skin, and gastro-intestinal symptoms.

Arsenic is a powerful drug and it seemed unnecessary to push it further than 30 or 35 minims in the day. If by that time it was not doing good it would be better to try some other drug.

Osler says he has often given as much as 25 minims three times a day but on the other hand he quotes a fatal case of arsenical neuritis in a child aged eight, who was given minims seven of Fowler's Solution three times a day for ten days. This was stopped for a week, then he took minims seven three times a day for a fortnight. He then developed neurtitis and he died.

Pigmentation/

Pigmentation of the skin may be produced by long continued doses of arsenic.

Most of the mild cases were given tonics particularly iron in some form. Before this is done attention must be paid to the stomach to see that it is healthy. Syr Ferri Phos. Co. or Cod Liver Oil and Malt in equal parts, were very useful for those cases that mainly wanted food and rest.

In those cases with violent movements chloral was the most valuable drug. This is perhaps best given as the Syr Chloral. It is well borne by children and is valuable if given frequently in severe cases. If there is trouble with swallowing it may have to be given per rectum.

The bromides dont seem of much use in the severe cases, but small doses of Pot Brom in combination with antipyrin are valuable in certain of the milder forms that attend as hospital out-patients.

For sleeplessness chloral is the best drug.

Six cases in this series were given chloretone in doses of grs 5 three times a day. This was given as recommended by Wynter, in ½ an oz. of petroleum emulsion. I cannot say that I saw any particular benefit from it, or encouragement to go on.

The/

The great object of rest in bed for even the mild cases is to try to stave off endocarditis and it is very difficult to know how long to keep those patients in bed. The very fact alone of rest in mild cases very often in a day or two practically stops the movements and if the patient is allowed to be about too soon the movements will probably return.

In some cases the movements persist for weeks and even months, but in some of those cases something can be done by suggestion.

If one sits by the bedside of such a patient and watches them, one can begin by trying to suggest that they keep quieter. This probably cannot be done, but in the next place if one grasp a hand and gently in addition try to control the movements, some little improvement may be noticed.
In the next place, if improvement continues certain voluntary movements may be tried under guidance. For example they may be directed to perform those movements that have been executed under control.

If those movements can be accomplished without spasms, then certain simple voluntary movements may be attempted without control.
All those movements must be of the very simplest character/

character, and their aim is to encourage freedom as well as precision in movement.

In most of the cases of chorea in pregnancy nothing special has to be done at all, and the pregnancy goes on to term. If the movements become very severe the question of emptying the uterus has to be considered.

According to Wall and Andrews mania in itself is not to be taken as an indication for this, but if this has to be done it ought to be done before the temperature gets high.

Many other drugs have been used and recommended by various observers in the treatment of chorea. For example Zinc and its compounds was much used by the older Physicians.

Wood recommends large doses of quinine, and antipyrin and cimicifuga have been much used in America. Ergot is strongly recommended by Eustace Smith, and strychnine by Ewart.

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