THESIS ON

SYDENHAM'S CHOREA

based on personal observation in

100 cases

— by —

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SYDENHAM'S CHOREA

Sydenham, who lived from 1624-1689 defined St. Vitus's Dance, as "a kind of convulsion, which attacks boys and girls from the tenth year to the time of puberty." As our present knowledge of the disease is chiefly theoretical, it is difficult to give a much more adequate definition than Sydenham did two centuries and a half ago. Holt says of chorea that it is a "functional nervous disease characterised by aimless, irregular movements of any or all the voluntary muscles. Choreic movements are of a somewhat spasmodic character, often accompanied by an apparent or real loss of power in the groups of muscles affected, and by a mental condition of extreme irritability.

Osler's definition of Sydenham's chorea is "a disease chiefly affecting children, characterized by irregular involuntary contraction of the muscles, a variable amount of psychical disturbances and a remarkable liability to acute endocarditis."

Brissaud defines it as follows "Chorea consists in purposeless and seemingly causeless involuntary movements occurring both during activity and rest, and consequently illogical and maladroit."
The salient points of Sydenham's Chorea then are, that it is essentially a disease of childhood, that it is characterised by aimless irregular movements of the voluntary muscles, and that it is frequently associated with acute endocarditis and psychical disturbance.

From the convulsive nature of the disease Sydenham's Chorea has been popularly labelled St. Vitus's Dance. The name St. Vitus's Dance was originally applied in Germany to a different disease, a disease which closely resembled the epidemic dancing mania which in Italy was called "Tarantism."

Hecker in his Epidemics of the Middle Ages states that "the Black Death had not yet subsided, and the graves of millions of its victims were scarcely closed when a strange delusion arose in Germany which took possession of the minds of men, and in spite of the divinity of our nature, turned away body and soul into the magic circle of hellish superstition. This "strange delusion" was characterized by a convulsion known as the dance of St. John or St. Vitus, on account of the leaping and frenzied dancing performed by its victims. This dancing disease seems to have been peculiarly infectious and it is stated to have been propagated by the sight of sufferers.

It was seen as early as 1374 in Aix-la-Chapelle,
whither it had come from Germany, and bands of men and women appeared in the streets, continued dancing for hours and appeared to have lost all control of their senses. The dancing took the form of wild delirium and was followed by great exhaustion. It is stated by ancient writers that when the disease was completely developed, the attack commenced with epileptic convulsions, and that those affected fell to the ground senseless panting and labouring for breath. They foamed at the mouth and suddenly springing up began their dancing amidst strange contortions.

In Strasburg in the year 1418 men and women began to dance and jump in the public market place. Many of them ate nothing for days and nights until their mania subsided. They were conducted on foot and in carriages to the chapels of St. Vitus, near Labern and Rokstein, where priests performed religious ceremonies on their behalf. Through the influence of St. Vitus a cure was expected, which was beyond the power of human skill. St. Vitus himself, was a Sicilian youth who suffered martyrdom at the persecution of Christians under Diocletian, 303 A.D. A legend was invented at the beginning of the fifteenth century that St. Vitus, just before his execution, had prayed to God that he might protect from the Dancing Mania all those who
should solemnize the day of his commemoration and fast upon its eve, and that a voice from heaven was then heard to say "Vitus, thy prayer is accepted."

Thus St. Vitus became the patron saint of those afflicted with the Dancing Mania, just as St. Martin of Tours, was at one time the succourer of victims of small pox. St. Anthony the friend of the erysipelatous, and St. Margaret the divine helper of puerperal women.

Many causes of the Dancing Mania have been mooted but into these it is unnecessary though interesting to enter, suffice it to say that at first it was considered as connected only with the moral and spiritual man, and not with his corpus, so much so in fact that for a century after its origin the treatment of disease was relegated by the physicians of the time to the domain of the church. In the beginning of the sixteenth century however, St. Vitus's Dance was made the subject of medical research, and ceased to be considered the work of evil spirits. Paracelsus was the man who was responsible for first directing the medical mind to the subject of St. Vitus's Dance. His sound common-sense, and practical mind are shown in his own words:

"We will not however admit that the saints have power to inflict diseases, and that these ought to be named after them; although many there are, who in their
theology, lay great stress on this supposition, ascribing them rather to God than to nature, which is idle talk. We dislike such nonsensical gossip, as is not supported by symptoms, but only by faith, a thing which is not human, whereon the gods themselves set no value."

Paracelsus divided St. Vitus's Dance into three groups:

- Chorea imaginativa (arising from the imagination)
- Chorea lasciva (arising from sensual desires)
- Chorea naturalis (arising from corporeal causes)

This last he explained by maintaining that in certain vessels, which are susceptible of an internal pruriency, and thence produce laughter, the blood is set in motion, in consequence of an alteration in the vital spirits, whereby involuntary fits of intoxicating joy, and a propensity to dance, are occasioned. One wonders if in this last variety Paracelsus is really describing Sydenham's Chorea. It was a much milder form than the other two, and did not differ so widely from our modern chorea. It is also interesting to note that whereas the chorea imaginativa and chorea lasciva were treated by appealing to the minds and emotions of the sufferers, the sickness of chorea naturalis were dosed with wonderful medicinal remedies.

The popular idea that chorea is propagated by
imitation is doubtless due to the fact that the Dancing Mania was undoubtedly so spread. Even as late as the 18th century Shetland was the scene of a nervous affection of a perfectly similar kind. It had its origin in a woman having an epileptic fit, while in church. Her contortions were soon imitated by many chiefly women but children also were affected. Samuel Hibbert, in his "Description of the Shetland Islands" tells how a worthy divine quelled the disease, by assuring his parishioners that the most effectual cure was immersion in cold water. As his church was situated near a small lake his proposed cure acted as a charm, and the devotions of the congregation were no more disturbed by the shrieks and writhings and tossings of the fair sex.

It is interesting to note that the Dancing Mania of Italy (tarantism) was supposed to be due to the bite of a peculiarly venomous spider, which rapidly diffused its poison through the body and resulted in violent convulsive movements. Madden in his "Illusions" however rather discredits this, as the convulsions did not usually appear until the year following the injection of the poison, so that we are left with the knowledge that the Dancing Mania was nothing more or less than a manifestation of hysteria, and thus an entirely different condition from our modern chorea.
Such is a short account of the true St. Vitus's Dance which has given its name in the popular mind, and even as a synonym in some modern standard textbooks to Sydenham's Chorea.

Having briefly discussed St. Vitus's Dance it would seem well to describe Sydenham's Chorea as Sydenham himself found it, before proceeding to examine more modern opinions on the subject.

Sydenham's definition of the disease I have already stated — a kind of convulsion which attacks boys and girls from the tenth year to the time of puberty.

"It first shows itself," he says, "by limping and unsteadiness in one of the legs, which the patient drags. The hand cannot be steady for a moment. It passes from one position to another by a convulsive movement, however much the patient may strive to the contrary. Before he can raise a cup to his lips, he makes as many gesticulations as a mountebank; since he does not move it in a straight line but has his hand drawn aside by spasms, until by some good fortune he brings it at last to his mouth. He then gulps it off at once, so suddenly and so greedily as to look as if he were trying to amuse the lookers on."

Such is Sydenham's graphic account of the symptoms of the disease into its aetiology he does not roam but
his treatment is worth quoting for its quaintness. It was as follows:

Bleed the arm to eight ounces, more or less according to age. Next day give half (more or less as the age of the patient requires it) of the common potion. At evening the following should be taken:

- Black cherry water
- Aqua epileptica Langii
- Venice treacle
- Liquid Laudanum

Make into a draught.

Repeat the cathartic every other day three times and the paregoric on the same nights. Blood must again be drawn the next day, and the catharsis repeated and so bleeding and purging must alternate until the third or fourth time, provided only that there be sufficient time between the alternate evacuations to ensure the patient against danger. To guard against a relapse bleed and purge for a few days that time next year or a little earlier.

He also prescribes mixtures of rue leaves, sage, betony, germander, white horehound, juniper berries &c. and adds that this treatment may also cure the epilepsy of adults, "but I have not tried."

As far as Sydenham’s treatment of the disease,
which bears his name, is concerned, it is true, that it differs widely from that of the modern physicians, but who is prepared to say that the modern Galen's methods are more efficacious in the treatment of chorea, than were those of Sydenham.

It is now my purpose to give a short account of the various theories as to the nature of chorea and its causation, to briefly describe its symptoms and diagnosis, and mention various lines of treatment, and to refer to a series of 100 cases from the records of the Royal Hospital for Sick Children, Edinburgh which I managed during my residency there as House Physician.
ETIOLOGY

The most prominent features of the general etiology of chorea are:

1. It is essentially a disease of childhood and adolescence.
2. It attacks females much more frequently than males.

The second and third hemi-decades provide most cases in fact Osler computes that three fourths of all cases occur then.

In the series of 100 cases which I have examined, and which are all cases of children under 13, the incidence in the various years is as follows:

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These cases comprise all the cases of chorea admitted to the Bathgate Ward of the Royal Hospital for Sick Children, Edinburgh, since 1898, so that in that time there has only been one case of chorea admitted to the ward under 4 years of age.

The Philadelphia Infirmary records include two cases entered as 'congenital chorea,' but Osler believes that all such cases are associated with definite cerebral changes and belong to an entirely different disease.

Sturges gives the proportion of females to males attacked as 3 to 1 and Osler as 2 to 1. My 100 cases yield 74 females and 26 males – almost 3 to 1.

Chorea is naturally held to be more common in the lower walks of life as statistics are particularly entirely drawn from hospital cases. While this is so, the mere fact of a child being reared in poverty exposed to neglect and hardship probably renders it more susceptible to chorea than its more fortunate brother or sister who lives in the lap of luxury, though he is by no means exempt from Sturges' "exaggerated fidgetiness."

Race. Weir Mitchell and Linkler have shown that the disease is rare, indeed almost unknown among negroes, and many have testified to the rarity of its occurrence among Indians.
In the "Transactions of the Association of American Physicians" in 1892 Morris J. Lewis published a paper entitled "A Study of the Seasonal Relations of Chorea and Rheumatism for a Period of Fifteen Years" and based his findings on an analysis of 717 cases.

His conclusions were to the effect that the seasonal incidence of chorea coincided with that of rheumatism, and that these tracings showed a marked resemblance to the tracings of the total amount of disease. It seems to me that not a great deal of importance can be attached to the fact that chorea flourishes at the same time as rheumatism does when concurrently disease in general has also reached its maximum. Chorea and rheumatism are not in this case twins, but only two members of a large family.

Lewis states that "the highest spring point of the chorea tracing corresponds with cool weather and a low barometer and mean relative humidity tracing, but the rise in autumn corresponds with cool weather and comparatively high barometer and mean relative humidity the temperature record therefore giving us but little information."
Seasonal Incidence of Chorea according to Lewis

Lewis has also arrived at the conclusion that the months of over study do not coincide with the months of greatest frequency of the disease.

To me it seems a well nigh impossible task to accurately arrive at the months of over study of over 700 children, far less to say that there are particular months when those 700 children are collectively prone to over study, or rather when all the children with whom these 700 are associated are liable or likely to overstudy.
The value of this conclusion of Lewis' is therefore surely very doubtful, and I think, he himself admits the charge when he states that "over study assuredly plays a most important role in predisposing children to chorea." Surely then if it were possible which I do not think it is to find the months of overstudy, one would find that these months were more productive of chorea than the rest of the year.

While chorea is a widespread disease, it is more common in towns than in the country. This however is doubtless due to the more healthy upbringing which country bred children are favoured with.

Osler has remarked that the disease tends to run in families and has found eighty cases in which other members of the family were attacked. Out of my 100 cases 7 show the occurrence of chorea in relatives, but only in two cases were those relatives directly related. Thus two children had brothers who had chorea, three had aunts similarly affected, one an uncle and one a cousin.

Jacobi believes that naso-pharangéal irritation is an important factor in the causation of chorea particularly of the face, but this is more likely to be a form of tic or habit spasm than a true chorea. Chorea verminaise was a name given by the old French writers to a chorea
caused by intestinal worms. Modern writers do not find a causal relationship between this condition and chorea.

Eye strain has also been mooted as a salient cause of chorea particularly by Stevens in the "New York Academy of Medicine Transactions," but though hypermetropia and astigmatism may produce habit spasm of the facial muscles there is no evidence that they produce true chorea. As Still says "there can be little doubt that much of the diversity of opinion concerning the etiology and pathology of chorea is due to the confounding together of various irregularities of movement which are in no way related to chorea."

Rheumatism and Chorea.

The place of rheumatism in the causation of chorea has given rise to much and lengthy discussion since it was first put forward by Bright in 1802. The same writer stated in 1820 that chorea is said to alternate with acute rheumatism "but through what organ or by what intervention it occurs is not conjectured." Many conjectures have since then been made, but the modus operandi of the diplococcus of rheumatism or of its toxins in the production of chorea has still to be discovered, nevertheless the fact remains that so many
'choreics' if I may so designate them have histories of rheumatism, that one cannot but believe there is some connection between the two ailments.

Statistics as to the frequency of the association of rheumatism and chorea cannot be relied on with too much confidence, as no uniform definition of what represents evidence of rheumatism can be laid down.

Of 100 cases I find that 13 had previously suffered from rheumatic fever or painful and swollen joints, while 3 of those 13 had rheumatic nodules, in two cases the elbows being the site and in the other the wrist.

Growing pains and pains in joints were complained of in 29 other cases.

In 7 cases a parent had had rheumatic fever, in 4 instances the father being the victim and in 3 the mother.

Statistics are plentiful to show how often chorea occurs in those who have suffered from rheumatism, but the exact percentage varies greatly and consequently it is needless to quote them, thus Steiner of Prague found only 4 instances of acute arthritis in 252 cases of chorea while English authors make it 20, 50 or even 70%.

The arthritis usually precedes the chorea, and but rarely is that order reversed, and Lee and Roger
have particularly insisted that chorea develops with the subsidence of arthritis, or may not follow until convalescence has been well established.

Though the clinician first drew attention to the association of chorea and rheumatism, the bacteriologist has also offered his contribution.

As long ago as 1872 Pianese claimed to have isolated a diplococcus and a diplobacillus from the cervical cord and cerebellum of a patient who died of chorea, and by inoculating cultures of these micro-organisms into animals he claims to have produced chorea. He states that the animals died "with muscular twitchings and convulsions," but it is open to discussion whether this was chorea.

In the Lancet of May 4th 1901 Poynton and Paine described the production of choreiform movements in a rabbit by intravenous inoculation of the diplococcus which they isolated in cases of acute rheumatism. Subsequent examination of the rabbit showed that the diplococci were present in the pia mater, and in the endo-thelial cells of the blood capillaries in the motor cortex. They also found micrococci in the motor cortex in a fatal case of chorea. Poynton suggests that affection of the pia mater is perhaps the important feature in the pathology of chorea.
Leonard Guthrie says:

Dana, Tarboulet Wasserman and Malkoff seem to have met with a similar diplococcus.

In the 'Lancet' (Sept. 22nd 1906) Dr. Andrews and Dr. Horder suggest that these organisms may have been evidence of terminal and incidental infections. Their efforts to isolate the diplococcus rheumaticus from the blood, pericardial fluid, and valvular vegetations in fatal cases of acute rheumatism were chiefly negative, and such as they did obtain they regarded as due to terminal infections.

In the New York Medical Record, March 28th 1908 Sachs then President of the New York Neurological Society has a paper entitled "Grave Chorea and its Relation to Septicaemia." In it he states that in one case of chorea in a woman of 22 he obtained cultures of Staphylococcus Aureus from the blood. This patient suffered from septicaemia and chorea, which was of an exceptionally violent nature, and the case terminated fatally. He also quotes other three fatal cases of chorea with marked symptoms of septicaemia in which no organism could be isolated. In only one of these three was an autopsy obtained and it showed the presence of verrucous endocarditis of the mitral valve.

Sachs considers that the chorea in the first case
was due to the Staphylococcus aureus and in the second to organisms not yet determined, adding that the search for a single organism as the cause of chorea seems fruitless and illogical, but that if we knew more of the elusive microbe of acute endocarditis we might be able to shed more light on the subject.

On studying his cases however one finds that the chorea was in evidence before the septicaemia so that, while doubtless the chorea was exaggerated by the presence of micro-organisms, we can by no means label them as the causal factor. The cases seem to be rather conditions of chorea with septicaemia superadded than real septicaemic chorea.

This experiment of Poynton and Paine supports the theory that chorea is due to the action on the nervous centres of blood vitiated by the rheumatic toxins. To this explanation of choreic symptoms there is the objection that as the blood is infected in every case of rheumatism, chorea should be much more frequent than it is.

In the B.M.J. of August 28th 1909 Macalister of Liverpool expresses doubt as to the frequent association of chorea and rheumatism, which he says rests upon certain clinical similarities and effects which may possibly result from an entirely independent
toxin. H. C. Ross has shown that if the leucocytes of healthy people are placed in the plasmata of other healthy people they live a somewhat shorter time than they do in their own plasma, whereas if they are placed in the plasmata of persons suffering from diseases which are due to, or associated with certain toxins in the blood, their lives are greatly shortened. From this he determined whether diseases of uncertain nature are accompanied by a toxin in the blood. By the use of this method Macalister has attempted to shed light on the relationship between chorea and rheumatism, and he came to the conclusions that there is in the blood plasmata of people suffering from chorea a poison which is toxic to the leucocytes of healthy persons, and that in the case of rheumatism the plasma appears to be hardly at all toxic to healthy leucocytes. He also found that the corpuscles of a chorea patient in the plasma of another chorea patient will live nearly as long as the corpuscles of a healthy person's will live in another healthy person's plasma and deduced from this that the choreic blood cells become immune to the toxin. If the toxin in acute rheumatism is the same as that in chorea the same evidence of immunity should exist when the corpuscles of a rheumatic patient are placed
in choreic plasma. When this was done, however, the lives of the leucocytes were invariably shortened thus showing that some difference exists between the persons in the two conditions.

And the difference in the blood of the two conditions found by McAlister is that choreic blood shows a marked eosinophilia while rheumatic blood does not.

The production of chorea by the vitiated blood of rheumatism cannot then be held to be proven, and I have tried to state above the present position of that question.

Another view which still holds rheumatism responsible for the production of chorea is that the choreic symptoms are due to a rheumatic affection of the heart - the rheumatic condition acting only indirectly.

Abnormal cardiac conditions certainly abound in chorea but again it has not been proved that they are the parents of chorea. Various opinions have been put forward as to the means by which the one condition leads to the other.

Bright thought the choreic symptoms resulted from irritation conveyed from an inflamed peridardium - or indeed pleura - by the phrenic nerve. This theory may hold good in those cases of chorea associated with pericarditis or pleurisy, but the number of cases in which
no such lesion exists is legion.

Kerkes, Hughlings-Jackson and Broadbent attributed the symptoms to endocarditis rather than to pericarditis and held that particles of fibrin from the inflamed valves produced embolisms of the minute cerebral arteries. That such an event is possible does not admit of doubt, but it is difficult to see why vessels of other parts of the brain should not be plugged as well as those of the motor ganglia. As Sir Dyce Duckworth says: "It would be truly remarkable if this embolic tendency was confined to the cerebral vessels only and equally common sites for such impactions, e.g. the spleen and kidney remained free, which indeed is the case. Again in vegetative endocarditis with embolism, one does not meet with chorea."

Dickinson in 1876 reported his findings in the autopsies of seven fatal cases of chorea. He found "hyperaemia, exudation and its consequences" but the cause of this state of affairs he could not find. As regards embolism, he remarked upon the absence of it, and upon the constancy with which the changes repeated themselves in certain positions, and the equality with which they affected both sides of the body as conclusive objections to the embolic theory.
Dana in 'Brain' 1890 describes the same vascular changes as did Dickinson, and Osler characterises them as being "the most constant central lesions in chorea." Osler reports one case in which initial endocarditis was very extensive, and in which was a spot of embolic softening the size of a cherry in the right lenticular nucleus.

The embolic theory is also at fault when one calls upon it to explain the great number of cases of chorea in which there is no endocarditis. But destructive criticism is easy!

Looking at the matter broadly, one sees that both rheumatism and chorea are diseases of the motor apparatus, the motor centres being attacked in the latter and the peripheral motor apparatus in the former.

Maclagan's ('Rheumatism,' by Maclagan) view is that the rheumatic diathesis implies liability to disturbance of the motor apparatus, and as the motor ganglia are an essential part of this apparatus, consequently the victims of rheumatism are liable to have motor ganglia which are more or less easily upset. He thinks that chorea caused by fright, shock &c. is only produced because of this susceptibility of the motor centres due to the presence of the
rheumatic diathesis, and that chorea may so arise in those who have had no definite attack of rheumatism.

This theory of Maclagan's is commendable in that it applies to all cases of chorea, but the exact modus operandi of the rheumatic diathesis is still to be found.

Whether then rheumatism produces chorea by rendering the motor centres susceptible or by producing endocarditis and embolism or by direct attack on the motor apparatus by its diplococci or indirectly by their toxins has still to be determined, but the fact remains that rheumatism and chorea occur so often in the same subject, as to suggest some connection between them, whatever that association may be.

With the exception of rheumatic fever there is no very definite relationship between chorea and the infective diseases, on the contrary the development of the acute exanthemata in the course of chorea usually check the disease. This is supported by Rilliez and Barthez, Radcliffe West and Trousseau.

There is also no evidence to show that any other disease save rheumatism is important in the production of chorea.
PSYCHICAL INFLUENCES AS THE CAUSE OF CHOREA.

There is no gainsaying the fact that the victims of chorea are almost invariably neurotic highly strung children. In 90 out of the 100 cases to which I have referred the nervous character of the children is specially referred to.

As Osler says, "bright-eyed, intelligent, active minded little girls, ambitious to do well at school, often stimulated in their efforts by teachers and parents, form a large contingent of the cases of chorea."

It seems to me that in these days of microbes and microscopes, too little stress is laid on this fact. Is there not an analogy between the maniacal insanity of adults and the chorea of children? Clouston states that the commonest post-mortem appearances in the brain in fatal cases of acute mania are intense hyperaemic conditions, while Dickinson, Dana and Osler state precisely the same fact with regard to chorea. Is it amiss then to call chorea a motor madness? It is at the age of 'co-ordination of motion and emotion' as Clouston aptly says, at a time when the motor apparatus is growing rapidly in knowledge and power, and has not yet grasped the nature of the duties required
of it by its owner that chorea most frequently occurs.

At this age motion is the means of expression most natural to a child, consequently when an emotional, highly sensitive child suffers either mentally or bodily, it is to be expected that he will give evidence of this by bringing his motor apparatus into play.

Fright, mental worry, excitement, great grief, &c. are undoubted frequent direct causes of chorea, and it may be as MacLagan suggests that the rheumatic diathesis predisposes the motor apparatus to be affected by those events, but it should not be lost sight of that those motor systems have belonged to people of neurotic temperament, which is defined by Leonard Guthrie as a "disposition in which the emotions are easily kindled, strongly felt and restrained or controlled with difficulty."

The dull phlegmatic man but rarely goes mad, nor yet does the unemotional staid child have chorea.

Sturges asks if "chorea may not be looked upon as a functional disorder, a motor disturbance which is as much beyond the reach of anatomical demonstration as are the various passions and emotions which, like it, distort and coerce the body in a great variety of ways?" He considers that the muscles concerned, having been educated more or less perfectly in certain
kinds of movement, are suddenly thrown back in their education by some nervous shock which enfeebles the capacity for stillness, which ought to develop pari passu with the acquisition of purposive movement. He also points out that the ataxy of chorea gives further evidence of the physical connection, being of a kind found in many conditions of mental excitement of perplexity and not of the pattern found in structural disease of the brain or cord, thus a choreic child may fail utterly in the performance of some muscular act one day, which next day it can accomplish with ease.

Sir Dyce Duckworth strongly believes that chorea is primarily a disease of the nervous system, and also that the susceptibility or instability of the nervous system is part of the rheumatic proclivity and given this state: an overt rheumatic attack, or some peripheral irritant - fright, or emotion, intestinal or utero-ovarian irritation may, he thinks, produce the peculiar perverted action of the motor centres.

On the other hand, Guthrie considers fright as shock under another name and is of opinion that its action in the production of chorea is by lowering vitality, enfeebling circulation and disordering metabolism, and so incapacitating the defending leucocytes, that the invading micro-organism, granting
their presence flourish and produce chorea. In addition, he says, that one must assume the presence of functional instability on the part of the nerve elements, otherwise chorea would be a much more common manifestation of rheumatism than it is.

The position then is that fright and other physical excitements, when acting on the possessors of unstable motor systems, undoubtedly contribute largely to the ranks of chorea.

It is suggested that these physical causes produce a merely functional disease, and it is on the other hand mooted that they produce an organismal disease, probably rheumatic, by weakening the defending phagocytes. Again, it is debated whether the motor instability of a neurotic temperament is in itself susceptible enough to be affected with chorea, or whether its instability must first be produced by the presence of the rheumatic diathesis.

Such then is the position of opinion with regard to the influence of physical and nervous conditions in the production of chorea.

Before leaving this subject, it should perhaps be said that among the most frequently operating mental causes of this malady is over-strain at school. Sturges particularly has remarked on this "school-
made chorea" as a serious injurious result of our modern methods of forced education. The frequency with which anxiety to excel at school intense application to lessons, &c., occur in the histories of children suffering from chorea, is certainly quite noteworthy.
Attempts have recently been made to class chorea as an infectious disease, but so far this hypothesis is not proven. Support, however, is lent to the suggestion by the fact that chorea sometimes becomes so prevalent as almost to amount to an epidemic. MaCalister, in the B.M.J., August 28th 1909, writes:—

"It has more than once happened in my experience that two children in the same family have developed the disease in succession, and on one occasion the return home of a convalescent child was followed by the admission of a sister with whom she slept. On making inquiries, I have sometimes found that two or more children in a school have had chorea in succession or simultaneously, and it seems possible that the old impression, that children got the disease by imitation may have arisen through the unsuspected infection of one patient from another."

I have already referred to MaCalister's experiments whereby he showed that there appears to be a toxin in the blood of choreics, and both Cabot and MaCalister have found eosinophiles marked in chorea, while MaCalister quotes a case of child who developed chorea in hospital, after occupying a bed next to a
chorea patient.

It is also urged in favour of the infectious nature of the disease, that while chorea endocarditis or articular diseases cannot be held in all cases to be productive of each other, yet so often are they associated that there may be a common toxin capable of producing them all.

Cases of chorea have been reported in conjunction with scarlet fever, gonorrhea and puerperal fever and such facts suggest the operation at least of infective processes.

Again the paralytic phenomena seen in some cases of chorea are suggested to be analogous to those occurring in typhoid and diphtheria.

This view that chorea is an infectious disease, however, does not explain those cases following fright and overstrain, unless of course Guthrie's view of the action of fright in lowering the vitality of the phagocytes and thus allowing the infectious agent to flourish is admitted to be correct.

Osler considers that cases of chorea, which occur in epidemic form and which older writers explained as being produced by imitation and which advocates of the infectious theory would explain by

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their's, are chiefly cases of hysteria.

They are probably more allied to the Dancing Manias of the Middle Ages. There is at least no evidence whatever that chorea can be produced by imitation.

PARATHYROID GLAND IN CHOREA.

In my study of the literature of the subject, I find that Limoire at the 6th Congress in Paediatrics held at Pavia in 1907 (Münchener Medizinische Wochenschrift, Feb. 9th 1909) stated that the removal of the Parathyroid gland in animals caused tremors and in-co-ordinated movements. He also records a case of chorea in which the administration of parathyroid gland gave very good results.

This of course is only of interest as an isolated case, and one cannot lay much stress upon it meanwhile. I only give it to complete my remarks on the etiology of chorea.
SYMPTOMS OF CHOREA.

Of the symptoms of chorea there is little now to be said.

The most marked and popular symptom is the spasmodic irregular movement, or 'exaggerated fidgetiness.' This may be mild, more severe or in the maniacal form, terribly pronounced.

The 'fidgets' are usually first seen in the hands and arms, which are also usually most severely affected, and may alone show the abnormal movements which may, however, spread to the face and legs and become general.

Chorea usually begins on the right side, may be confined to one side of the body - hemichorea - but this was the case in only one of my 100 cases.

Landon-Carter Gray of New York, writing in the fourth volume of the 'International Clinics' lays great stress on the fibrillary character of the muscular movement and also on its sudden beginning and equally sudden ending. He thinks that this can be felt even when it cannot be seen by taking the affected limb in the hand and recognising the fibrillary muscular contractions and he goes so far as to diagnose the presence of chorea from these
fibrillary twitchings which he can feel but cannot see.

The essential point about the movements is that they are irregular and involuntary.

They cease during sleep usually but not in every case.

In practically all cases of chorea emotional disturbance is marked. The child is keenly sensitive, indeed the characteristic movements are often in abeyance until the child is conscious of being observed. Crying is a common occurrence, and fear and terror are also frequently marked.

Premonitory symptoms are sometimes present in the shape of headache, digestive disturbances and general restlessness.

In nearly one fourth of the cases the speech is affected, due more to the irregular movements of the lips and tongue than to affection of the laryngeal muscles, the impairment being thus rather in phonation than in articulation, and sometimes but rarely this difficulty in speech is the first symptom noted.

The gastro-intestinal muscles, the bladder and muscles of the bronchi are never involved, and though the irregularity of the heart so common in chorea is by some attributed to spasmodic contraction of the
heart muscle, there is no evidence to show that this is so.

The severity of the symptoms of chorea depends on the type of chorea, of which Osler notes three - mild, severe and chorea insaniens.

The affection of the facial muscles gives rise to the production of grimacing, and the child often appears to be expressing various emotions - such as joy, fear, dislike, surprise, &c. Movements of the jaw, tongue, and lips produce often a peculiar 'clicking' noise.

Pupillary changes also occur. In the Lancet of Jan. 18th, 1908, Dr. Langmead described hippus as sometimes occurring, also inequality and eccentricity of the pupils. Irregularity in the shape of the pupils may also be seen, and nystagmus is sometimes present.

Braxton Hicks in the Lancet of 1889 reported a case of chorea in pregnancy where the uterine contractions became very irregular.

Quite frequently there is enfeeblement of the muscular power, but according to Osler rarely true paralysis. This muscular weakness may be hemiplegic or paraplegic but is most frequently monoplegic. This loss of power usually comes on after the choric
movements have made themselves evident but it has been known to occur before then. It generally disappears with the cessation of the movements, but wrist drop has been known to persist for two years after an attack of chorea.

In severe chorea loss of weight may be marked, and even if only one limb is affected it may show marked atrophy.

Sensory symptoms in chorea are not pronounced, but sometimes there is pain in the affected limb or limbs, even although there is no arthritic affection.

Some observers have also found pain situated over the points of emergence of the spinal nerves but this is by no means a common phenomenon.

Headache is the most frequent painful sensation accompanying chorea, and sometimes it is present as a premonitory symptom.

The knee-jerks in chorea do not usually differ much from the normal in intensity, if anything they tend to be diminished.

In the British Medical Journal of March 30th 1901 Dr. W. Gordon referred to a peculiar modification of the knee jerk which he found very commonly in chorea. He described it as follows:

"With the patient recumbent if one raises the
knee, allowing the heel to rest on the couch, making sure that all the muscles of the limbs are relaxed, and if one then tests the knee jerk in the usual way the foot is found to rise more or less smartly, but instead of falling back immediately, it remains suspended for a variable time—hung up as it were—and then slowly sinks back to its original position.

There are also variations. Sometimes the peculiarity amounts merely to a sluggish descent, following an ordinary ascent; sometimes an ordinary knee-jerk is obtained, but just as the foot is beginning to fall again, it is caught in mid-air and held for a time, or even raised to a higher level than that reached in the first jerk. Sometimes the knee jerk passes at once into an active, more or less persistent, rigid extension of the limb. And between all these varieties there is every gradation.

So far as I can find in the literature of the subject this is the only reference I can find to a peculiarity of the knee-jerks.

A careful examination of the records of the 100 cases to which I have referred, however, shows the somewhat astonishing fact that in 12 of them the knee-jerks are described as being not normal or diminished, but exaggerated. As all these cases
are not reported by the same resident physician, it seems probable that in some at least of them this was really so.

Again, I find a case of a girl of 8 years whose knee-jerks at the beginning of her illness were absent while after the chorea had lasted 2 months (it lasted almost 6 months altogether) the knee-jerks were exaggerated.

This would seem to suggest that there is no uniformity in the knee-jerks in chorea.

Psychical disturbance has been already referred to, but the special senses are rarely involved. In maniacal chorea however hallucinations and delusions may be marked, and melancholia has been described. Maniacal delirium occurs in chorea insaniens, and the existence of the chorea has been overlooked, and the sufferers removed to asylums.

Chorea insaniens is fortunately rare and Osler has only seen two cases. He compares it with regard to chorea as he would malign scarlet fever to the ordinary scarlatina.

He describes it as usually occurring in young females between 15 and 20 years, but sometimes
attacking children and quotes a case in a girl of
nine years where death occurred in 130 hours. While
chorea insaniens may begin as such, it usually com-
mences as an ordinary chorea, and gradually develops
with hallucinations delirium, - sometimes mania. The
temperature is nearly always raised and may be very
high (104-107°) and it is noteworthy that the move-
ments may diminish or even cease before death. The
duration of fatal cases is rarely more than 2 weeks.

All my 100 cases were cases of ordinary, mild or
severe chorea, no case of chorea insaniens occurring
among them.

Fever, which falls to be noted in the maniacal
form is not characteristic of the ordinary case, and
if present in such is probably due to some complica-
tion - acute rheumatism, endo- or pericarditis.

Cutaneous affections in chorea chiefly relate to
pigmentation from the therapeutic use of arsenic, or
to rheumatic affections - erythema nodosum or purpura.

In 100 cases, I only find 1 noted as having
purpura, while 3 showed the presence of subcutaneous
rheumatic nodules - 2 in the elbows, and 1 in the
wrist.

With regard to the blood reference has already
been made to Macalister's finding of the probable
presence of a toxin in the blood of chorea patients while he has also found eosinophilia present in which latter respect he corroborates Cabot. Various observers have found different organisms present in the blood of chorea patients, but so far there is no unanimity of opinion either as to their origin or effect.

The urine shows no constant abnormality. Increase in the output of urea, of phosphates of albumen and of sugar have all been described, while uric acid has also been found increased and the chlorides diminished.

The presence of uro-haematoporphyrin has also been noted both in chorea and rheumatism and this fact has been put forward as showing some connection between the two maladies.

Several observers have had cases wherein acute nephritis developed, but this cannot be considered as a common complication.

Heart Symptoms in Chorea.

Heart symptoms, or perhaps more correctly, signs of disturbance of the heart's action and mechanism are very frequent in chorea.

The most common abnormal cardiac conditions found
in this disease are endocarditis and pericarditis and chiefly the former.

Pain about the heart is rare, and so is palpitation. Irregularity of the heart's action is sometimes present and by some this has been held to be due to a choreiform spasm of the heart muscle. This view may be taken to be now abandoned for if it were true the irregularity would surely be much more frequently present than it is, and Osler remarks that he has never seen a case where the irregularity was of such a kind as could be attributed to choric action of the heart muscle.

Another theory is that the irregularity is due to disturbance of the respiratory movements of the thorax, and of the two this seems the more feasible.

A much more common form of abnormality in the heart's action is rapidity, and this is only to be expected, when one remembers that the subjects of chorea are prone to outbursts of emotion.

A still more common and more important condition however, is the presence of heart murmurs, which may be either functional or organic.

The production of a functional bruit in chorea is aided by the frequent presence of anaemia and debility and corresponds to the similar condition
found in debility from fever, and neurasthenia, &c.

Such a functional bruit usually consists of a soft systolic murmur most commonly of greatest intensity in the pulmonic area, but sometimes to be found in the aortic region and also at the apex. With such murmurs a diffuse apex beat and venous pulsation in the neck are commonly associated.

Walshe thought that these murmurs were often caused by disordered action of the muscular apparatus connected with the valves, — a want of correspondence between the fibres of the ventricle which obliterate the cavity and those which close the valve. This is experimental and conclusive evidence to hold this theory. Dr. Townsend Porter, of the Harvard Physiological Laboratory by ligation of the ramus descendens of the coronary artery in animals produced infarction of the anterior papillary muscles from which the chordae tendineae pass to one half of each segment of the mitral valve, and on auscultation after complete infarction no bruit was heard.

While those functional bruits are common they are no more so than are true organic bruits in chorea, which of all diseases is most frequently associated with endocarditis, indeed in the records of post mortem examinations of fatal cases of chorea, it is
rare to find mention of the heart being free from endocarditis.

Frequently without clinical symptoms or signs during life, valvular endocarditis has been found post mortem when by far the most common form found is the presence of small warty vegetations on the auricular surface of the mitral valve. It has been suggested that the bruit is produced by the friction of the diseased segments on each other and not by regurgitation, hence the lack of symptoms. This view is supported by the fact that the bruit often disappears, which may be accounted for by the contraction of the vegetations and by their becoming smoother and firmer. But still the effects of the endocarditis do not differ from those of ordinary endocarditis, thus Osler who re-examined 140 cases from 2 to 16 years after they had suffered from chorea found definite and persistent damage in \( \frac{51}{2} \% \) of them. This destroys Sturges' statement that "none of the injurious after consequences which attend endocarditis in its other relations are found in chorea;" and is conclusive evidence that the endocarditis of chorea is responsible for the production of organic heart disease.

The most common heart lesion then in chorea is the presence of warty growths on the segments of the
mitral valve produced a systolic murmur such as is obtained in mitral insufficiency.

In the reports of 100 cases I find the presence of heart murmurs noted in 42 of them, and of these 30 were systolic bruits in the mitral region, 8 giving a double mitral murmur, and 2 aortic and pulmonary systolic murmurs, while 2 showed mitral presystolic murmurs.

Pericarditis is also an occasional complication being found in 19 or 73 autopsies by Osler. I do not find it noted in any one of 100 cases.

In those 100 cases I find it noted in 20 that reduplication of the pulmonary 2nd sound was present, in one of which all the sounds were reduplicated. In them too accentuation of the pulmonary 2nd sound was fairly common occurring in 15.

The causal factor of endocarditis in choreas is unknown. In some cases acute rheumatism is responsible but in cases where there is no history of rheumatism, the physical signs of endocarditis are often found, and of the two diseases, chorea and rheumatism, the former is now considered to be more productive of endocarditis than the latter. There are those who believe that chorea, rheumatism and endocarditis are all produced by one and the same
organism but so far this almost omnipotent microbe has not permitted itself to be demonstrated.

Before concluding these few remarks on the symptoms of chorea, I must make reference to a paper entitled 'Latent Chorea' by Dr. Miller of Paddington Green Children's Hospital, published in the B.M.J., of Dec. 18th 1909.

Dr. Miller describes 'Latent Chorea' as a particular condition of mental and physical ill-health which is rheumatic in origin, and which may be seen in children with obvious acute rheumatism in children convalescent from rheumatism and chorea and in children without any very obvious rheumatic symptoms.

He considers rheumatism and chorea to be part of the same infection, and thinks that 'latent' chorea can frequently be seen in nearly all children with acute rheumatism, who do not show signs of overt chorea. He regards as evidence of 'latent' chorea in such cases fidgets, clumsiness and the child's inability to keep its eyes fixed on a particular object for more than a minute or two. Mental depression common in acute rheumatism and frequently regarded as due to treatment by salicylates, Dr. Miller
regards in many cases as evidence of chorea lying dormant or about to develop.

The signs of latent chorea in those convalescent from acute rheumatism, he thinks, are mainly mental, chiefly mental exaltation following morbid depression, and this he considers is present alike in children convalescent from acute rheumatism, rheumatic synovitis and chorea.

In those with no symptoms of rheumatism Dr. Miller still find evidence of latent chorea in headache, languor and loss of appetite, extreme nervousness and timidity, fidgetiness and irritability and clumsiness.

He also finds that such children often become stupid at school, although previously doing excellently there. Talking during sleep, somnambulism, difficulty in getting to sleep, enteric diarrhoea, nocturnal enuresis he states as the result of nervous instability produced by such chorea.

Dr. Miller concludes that chorea declares itself first by symptoms significant of general nervous instability, and that the well known association between rheumatism and nervous instability is due to the latter being caused by latent chorea, and that the presence of latent chorea in children suffering from
acute rheumatism affords strong evidence that chorea is a rheumatic condition.

While Dr. Miller's paper is of undoubted value in pleading for an earlier and more extensive recognition of chorea, the symptoms he gives as being evidence of latent chorea appear to me to be such symptoms as may be found in well-nigh every neurotic and highly strung child, who shows evidence of mental instability.

He has not shown reason to deny that this nervous instability is not as likely to produce chorea as vice versa. To my mind in describing 'latent chorea' Dr. Miller has described the condition of children from whom the ranks of true chorea minor are almost entirely recruited.

With regard to the 'Electrical Reactions in Chorea', Gowers, Schmitt, Rosenthal and Benedikt agree that in some cases there is increased electrical instability both in nerves and muscles, and to both galvanism and faradism and this is not constant.
DIAGNOSIS.

Diagnosis of a well marked case of typical chorea presents no difficulty, the peculiarly characteristic form of the movements being unmistakable.

In chorea insaniens the true nature of the disease may be overlooked, and the case classed as one of acute mania. Acute mania, however, is exceptionally rare in young children, among whom chorea is chiefly seen and some few typical choreiform movements may appear to give one the clue that the case is one of chorea, though the movements are often in abeyance in chorea insaniens.

Hysteria may simulate chorea closely, but the hysterical movements are more rhythmical, and imitation may be found to bulk largely in the production of hysterical chorea, but not of true chorea.

Friedrich's ataxia is distinguished by its hereditary character, the scanning speech, and the slow inco-ordinate character of the movements.

The spasmodic movements of cerebral disease in infancy may simulate those of chorea closely, but in such gross cerebral disease one finds defective intelligence, rigidity, while the onset of the
disease is in infancy and its course is chronic.

Paresis from chorea especially when the typical movements are absent as they may be, may be confused with poliomyelitis or paraplegia, but in chorea there is an absence of atrophy and of the reaction of degeneration, and the spasmodic movements are extremely rarely entirely absent.

Chorea and Tic.

The movements of chorea are involuntary and not amenable to control by the patient, while those of tic are always volitional, if not voluntary, and may always for a short time at least, be controlled by the will.

In chorea the movements are inco-ordinate and purposeless, and as Professor Patrick of Chicago, in his paper on Tic and Chorea (Journal of American Medical Association, May 1st 1909) says pathologic in the association and sequence of muscular contraction, while those of tic are co-ordinate and purposive, and physiologic in the association and sequence of muscular contraction.
PROGNOSIS.

The immediate prognosis of chorea is distinctly good. In 100 cases I do not find a single death. The average duration is about 6 weeks, but in one of my cases the choreic movements lasted for 5 months.

According to Osler the very chronic cases - 6 months or more - are usually forms of tic, though he quotes a case which had definite chorea for 3 years and then recovered.

Recurrence is very common particularly in the Spring, and statistics go to show that almost half the cases of chorea have more than one attack, and some have been described as having had six.

Osler considers a mortality of 2% a reliable estimate.

With regard to the sequelae the only one which is really persistent is endocarditis. Mental dullness and paresis of limbs usually pass off.
TREATMENT.

There is no difference of opinion with regard to the general principles of treatment of Sydenham's chorea, and these may be summed up as first and foremost rest and sleep, secondly abundant and nutritious food. By far the most important item in the treatment of chorea is rest, indeed rest and feeding was all the treatment offered to the chorea cases in the Bathgate Ward of the Royal Hospital for sick children, Edinburgh, during my residency and all the cases did thoroughly well.

The seclusion of the patient by screens round the bed in hospital, or by isolation in a perfectly quiet room at home, assist the repose of the child greatly and not only this, but such seclusion is doubtless beneficial in allaying the fears and timidity of the keenly hypersensitive and emotional children, who as we have seen are the usual victims of chorea.

In severer cases it may be necessary to protect the child from injuring itself by a suitable arrangement of bolsters and pillows, and for this reason also the parts of the body most likely to be contused may be padded with advantage, e.g., the knees and elbows. Where the spasmodic contractions are very severe, it
is better to have the bed made up on the floor, to prevent injury to the child by falling or rather being thrown out of bed.

The diet should be nutritious and easily absorbed, e.g., milk, eggs, and light soups. The more food the patient can take the better. In severe cases where deglutition is difficult it may be necessary to resort to nasal feeding.

With regard to the drug treatment of chorea, arsenic is undoubtedly the most generally used remedy, although no drug can be considered as being a specific in the disease. Arsenic is not held to have any specific action, but probably is advantageous on account of the improvement it produces in the general nutrition. Fowler's solution is a convenient form in which to prescribe it and the dose to start with should be small. Three minims after meals well diluted. This may be increased by one minim per dose daily until the child is taking 15 minims thrice daily, and the good effect of the drug is not usually seen until the maximum dose is taken. Dr. Mauray of Newcastle recommended large doses (15-20 minims) from the beginning but admitted that this method if persisted in too long is not devoid of risk.
Osler does not think that arsenic has any effect on controlling the choreic movements. When large doses are given, an careful watch must be kept for symptoms of poisoning by the drug, e.g., vomiting, diarrhoea, itching of the skin, oedema and pigmentation of the skin. The prolonged administration of arsenic in large doses may also induce peripheral neuritis.

Dana recommends 5 drops of Fowler's solution dissolved in half a teaspoonful of the syrup of the iodide of iron or accompanied by a grain of the citrate of iron and quinine in solution the dose of the Fowler's solution being increased to ten minims three times a day and occasionally more.

Dana also got good results from exalgine given up to 3 grains five times a day with one grain of iron and quinine citrate.

Zinc compounds were a popular remedy at one time and still are with some. In the 'International Clinics' published in 1894 Dana states that he has seen great benefit from the administration of Zinc Bromide.

To control the movements chloral hydrate is recommended, particularly by Dyce Duckworth ('International Clinics, 1891), but sleep induced
by chloral hydrate is liable to be followed by maniacal excitement.

Hughlings-Jackson advised the administration of alcohol - a teaspoonful of brandy every four hours - and he got excellent results ("Braithwaite's Retrospect of Medicine", vol. XVIII.)

Among other remedies antipyrin, cimicifuga, sulphonal and physostigmine have all been recommended as also has quinine.

The salicylates have not proved of much benefit even in those cases of chorea associated with arthritis. Strychnine, belladonna, silver nitrate and bromide of potassium have also their advocates.

Massage, electricity and gymnastics have been found useful in the convalescent state and change of air and scene are often very beneficial in chronic cases.

According to Ollerenshaw, Isopral is a useful drug to allay the movement in chorea, and Knop and Fabian report two cases where the injection of atoxyl produced an apparently permanent cure.

Most recent of all the remedies advocated is the injection of sea water under the skin covering the scapula.

As I have already stated, Simoni records a case in which the administration of parathyroid gland gave very good results.
In 'Current Medical Literature' vol. 52, No. 17 Lees recommends that the essential factor in the treatment of chorea is to put the child to bed and treat it as a case of rheumatism, and he would give 400-500 grs. of sodium salicylate per diem. This has not been found successful by Dr. Voelcker.

Chloretone Promural and Trional have all their adherents and Baccelli of Rome urges the use of Monobromate of Camphor (15 grs. daily pushed to a maximum of 30 grs. daily). Among the hypnotics, hyoscine is also largely recommended.

Thus it may be seen that while many drugs are called upon in the treatment of Sydenham's chorea, few are chosen as being reliable, indeed one may say that none are, so that as far as the medicinal treatment of the disease goes, it is doubtful if much advance has been made on Sydenham's Black Cherry Water, Venice Treacle, rue leaves, betony, germander, &c.

The essential point in the treatment is to keep the child at rest and isolated though this last Koplik does not believe in.

One word with regard to the preventive treatment of chorea and I have done.

As such Sydenham recommended bleeding in the Spring following an attack, but surely of much more
importance is the recognition of the neurotic temperament in children. These children are undoubtedly predisposed to chorea on account of the instability of their nervous systems - if such children were guarded from over-strain at school, as much as possible from terror and were intelligently understood by parents and teachers as being different from their more staid and perhaps more uninteresting brothers and sisters, then it is highly probable that Sydenham's chorea would occur less frequently than it does.