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A CONTRIBUTION TO THE ETIOLOGY OF  
CHOREA.<sup>1</sup>

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THE mechanism that prompts the peculiar, incoördinate movements which make up the objective neuro-muscular phenomena of chorea has never been explained, and is to-day as much a mystery as in the time of Sydenham.

The recorded pathological evidences serve only to prove some irritating process has been at work upon the cerebral and spinal motor tracts and coördinating adjuncts.

Though meagre, and, in the main, unsatisfactory as aids in the solution of the causes of chorea, still these pathological lesions help, with other associated factors, to adjust certain etiological sequences. It is my endeavor in this communication to bring to bear united and associated pathological and clinical experiences, in the adjustment of certain facts, to better the understanding of the nature of chorea.

The mortality from chorea seems to be greater in

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<sup>1</sup> Read before the New York Neurological Society, December 6, 1892.

Great Britain, and the English observers seem to have given more attention to the gross lesions (which apparently are the result of complications), which chiefly are cardiac valvular troubles and emboli, pericarditis and vaso-motor changes.

The reliable accumulated data of the microscopical conditions of the brain and cord is somewhat limited. It, however, attests that some morbid irritant has been at work.

The epitomizing of these evidences has been admirably done by Dr. Dana in his summary of the consideration of thirty-nine cases, up to 1889, viz. :

“ We find that in the sub-acute types of chorea there is a hyperæmia of the brain and parts of the cord.

“ In the brain this is not meningeal but subcortical and basal. The arterial walls are paralyzed, dilated and badly nourished, so that exudations occur and the lymph spaces become distended and eroded. There is sometimes stasis, thromboses and spots of softening, or the walls give way; there are minute hæmorrhages. The lymph spaces around the ganglion cells are not dilated. In other cases the vascular and neuro-degenerative changes are marked. The small arteries are permanently dilated, a little thickened and degenerated. Peri-vascular channels may be more eroded and distended. There is now some connective tissue proliferation and signs of degeneration in the ganglion cells. The nerve fibres show varicosity. *Hyaline bodies are seen.*<sup>2</sup> In fine, we have in chorea, first, vaso-motor paralysis and trophic disturbances, affecting certain areas of the brain, and, to a less extent, the cord. Then we have—this becoming chronic with connective tissue—hyperplasia and degenerative change in ganglion cells and fibres.”

Debarring the grosser concomitant pathological features, such as emboli, extended softenings, etc., this *résumé* tells the story as accurately as it is possible to be told.

It begins in hyperæmia, results in exudations, is complicated with hyperplasia of connective tissue elements, and the action takes place throughout the motor areas, and the end is resolution.

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<sup>2</sup> Italics are mine.

The degree of hyperæmia and the amount of exudation of lymph, and its products, are relative. The overgrowth of the connective tissue in mild cases is hypothetical. The morbidic stimuli, which incites the nutrient distribution to the motor-areas to rebellion and disease, must in some way, by natural selection, choose this location. There is as well-defined predilection shown through the nutrients for this widely-distributed area as is shown in rheumatism of adults in the selection of the joints of the extremities.

What is this morbidic factor with its special predilections?

We perforce are obliged to seek our information from clinical study, and from experience make up our deductions from logical inferences and analogies solely.

The following case I shall use as a text for the consideration of some of the more prominent reputed causes of chorea:

H. H. S. (eleven years of age).—Came under my care at the Presbyterian Dispensary early last June. He had been in the hospital about two weeks, and was transferred to my department for treatment.

*Family History.*—Careful questioning of the mother developed very little of interest in this direction, except rheumatism was in the family on both sides, and the mother was neuropathic.

*Past History.*—The boy was always delicate, nervous, and of an irascible temperament, and very difficult to govern; attacks of indigestion were frequent, and sore throat. Two years ago he fell from the top of the stairs of an elevated road station outside of the railing to the street. The fall was prompted by fright. The porter suddenly appearing, and threatening the boy, his presence of mind fled, and he fell. He was stunned and carried home half unconscious. No especial injury was noted except the breaking of the two front upper teeth. Six months after this first appeared choreic movements of face and upper extremities, with sallow complexion, and some complaints of pain in back and limbs. Chorea grew more pronounced and became general. The attack lasted about ten weeks. Another attack came on in a year, slight and of shorter duration. He wet the bed

nights, and, when finding himself damp, he would jump up, take off his night clothing, and return to bed, and would be found cold, shaking, and teeth chattering, etc.

*Present History.*—About eight weeks prior to the boy's entering the hospital, he complained of erratic pains throughout the body, back, thighs, knees, arms and head; worse at night, and severe enough to keep him awake and cry. No rise of temperature noted, or swelling of any joints. Very soon small tumors made their appearance on the smaller joints of the extremities—little soft lumps, capable of being rolled under the skin. Rapidly, day by day, increasing in number and extending up the extremities, along the muscles to the knee and elbow joints—most entirely confined to the extensor surface. Then, in about six weeks (two weeks prior to entering the hospital), he commenced to have another attack of chorea, which in a few days was very pronounced, and in conjunction with the attack he complained more bitterly of pains, and locomotion was difficult, and swelling of ankles and wrists noted. Pains were worse on movement. He suffered on going upstairs; tired easily; was very pale; listless; water less free, and skin harsh and dry; no fever noted; the tumors increased in number.

*Examination.*—Anæmic, richitic chested, undersized, thin, dry, harsh skin, enlarged smaller joints, marked puffiness of ankles, wrists and knees, all tender on movement, choreic movements general, but not extremely severe, speech affected slightly, hoarseness of voice, pot-bellied, diffused heart impulses, exaggerated action plainly visible, mitral regurgitant murmur, heart enlarged, irregular in rhythm and force.

The boy was plentifully covered with about one hundred and fifty (actual count) little tumors from occiput to the extreme smaller joints of the toes. They varied in size from the head of an ordinary pin to a good-sized almond. There was only one of this size, however, situated near the r. olecranon process. They were more frequently about the size of a small pea. They varied somewhat in consistency. None, however, were very hard; some were firm, yet most of them were soft; the smaller ones gave the impression of being the hardest. There were a number which felt as though they were semi-solid, and the large one felt somewhat like a ganglion so often seen on the tendons of the extensors of the hand, in a day growing to full

size, and growing smaller in a week, hardening down in some cases, and others remaining soft as they grew smaller. At first many of these were double the size than when first seen by me. The house physician informed me he had excised a part of one and microscopically it revealed young granulating connective tissue, very soft in texture, and lacey.



Their anatomical position varied; some were immediately associated with the skin; others were below it in the connective tissue, rolling freely on manipulation; others seemed attached to the sheaths of the tendons of the muscles; some seemed to be next to the periosteum. One on the occiput was not movable, quite dense in structure and was apparently attached or connected to the periosteum.

In my endeavor to extricate a small superficial soft one on the right forearm, it broke and entirely disappeared. The mother refused to allow another being extracted.

The photograph gives a fair representation of the body distribution and size of these little nodules. The further discussion of these nodules I will leave to its place in my paper. These nodules varied in their permanency, some lasted only a week or two, others a month; others two and three months.

The boy's urine was 1023 specific gravity and acid in reaction. The blood showed a diminution of the normal number of red corpuscles.

There has been no treatment. The heart has improved, both in diminution of murmurs of irregular rhythm and force. The chorea has not ceased; nutrition has improved. The rheumatic symptoms are not yet silenced. The boy still has nocturnal enuresis, and the specific gravity of the urine is 1021. The blood microscopically appears normal.

It will, no doubt, be agreed that we have here an interesting case.

If I should read a hundred histories, there could not be selected one which could be more instructive, nor would the combined statistical information or the clinical composite photograph vary in the general outlines.

One interesting factor must be noted, no matter what the chief reputed etiological disturbing factor in chorea may be,—heredity shock (emotion or nervous tension), rheumatism, reflex causes, infection or pregnancy,—the phenomena of the disorder remains the same. The picture is, except in degrees of severity, always positive.

There is a uniformity in the objective neuro-muscular phenomena which never necessitates long chapters on differential diagnosis; but conjoined with this there are physical addenda too often slighted. Many of these are combined in the case related; a badly nourished child, anæmic, rachetic; subacute rheumatism; unsound heart. A general *abortive* picture from the health line.

What is directly to blame for this very often observed complex symptom designated subacute chorea?

Let us consider briefly some prominent reputed etiological factors.

Among the chief predisposing ones is *heredity*. It bears, no doubt, equal importance and relationship to all systemic affections and diatheses.

A relationship in the sense that a disposition is sown by diseased parentage, which results in some deviation to normal resistance to any severe systemic disturbance. A vice of constitution, direct or indirect.

Permit me here to quote a rather unique conceit in reference to this point, from the pen of Dr. Collins :

“ For instance, we may see running through whole families, certain peculiarities, which, at first sight, might not show any inter-relationship, but which, on close study, are shown to be but pollen from the same anthers, that has disseminated itself by marriage and propagation through many members of related families.”

Heredity is freakish. Heredity is such a broad subject, and surrounded by so much speculative and uncertain mysticism, that no direct information is attainable in half the cases we study. Positive information is rarely obtained. There is no more advance in this line of study than was known a century ago.

*Age*—Is an important positive and relative factor.

As a rule, chorea is confined to the pre-developing period of man—and womanhood. The exceptions serve to prove the rule. They are few and irregular, often in the picturings. Thus, anomalies are often hysterical. One, in observing senile chorea, feels bound to believe in revision of type in the picture of this second childhood.

Age bears an importance to chorea when it is compared to correlative and associated diseases.

Rheumatism, heart diseases and the subcutaneous nodules.

Age bears to these exactly the same relation.

Logically, in this relationship, chorea bears some association and a common etiology with them.

*Sex and Pregnancy.*—These may be considered together. The preponderance of the female of three to one, is quite uniformly observed. Its being also associated in certain cases of pregnancy naturally raises the question, Why this sexual affectability?

Laycock, Denis and LeCarne, all lay stress upon the especial condition which determines sex influences to the vascular system.

Concisely, this is a condition of more water and less crassementum, a so-called looser crases of the blood. It is universally considered that the female's nervous system generally sooner responds to all morbid stimuli.

The term affectability, applied by Morell to the constitution of woman, is expressive. She is more apt to be afflicted with certain affections.

This predisposition or affectability is shown as well for rheumatism, endocarditis and subcutaneous nodules. As man grows older, past the age of puberty, he grows less predisposed to chorea and allied troubles—especially to chorea and the subcutaneous nodules. This is not so with woman. The vasomotor element and her affectability continues on for a longer period, and way past the menstrual line. There is apt to be an maxiosed affectability in deranged menstruation—unfortunate pregnancies and prolonged nervous tensions.

This applies equally as well to rheumatism—cardiac troubles, subcutaneous nodules as to chorea.

Anæmia and mal-nutrition are causes which directly apply and equally to all correlative affections of chorea. These need not be dwelt upon further here.

*Shock.*—All children in their history of development are subject to mental excitement.

Sudden frights are common. The effects, immediately apparent, are vasomotor disturbances, especially upon the heart. Pallor is seen and more or less reflex emotional disturbance, and more or less facial muscular excitation. The direct effect, however, is upon the heart, and unless this be in some way unhealthy is not of long duration. Slight effects upon higher nervous centres.

and through vasomotor disturbances upon nutrition is granted; but why it should affect the motor ureas so extensively as to produce chorea I do not understand.

Chorea, as far as I have investigated, is rarely immediately induced by sudden shock.

My experience relates only to two cases. The shock occurring one day and followed by choreic movements the next. The one was a primary attack followed in a year by another attack, accompanying rheumatic phenomena. The other case was not followed up for any length of time.

I do not believe in sudden shock as such a prevalent exciting factor as is credited to it by so many writers and the laity.

Statistics upon this point, to my mind, are not reliable. I consider it a very rare exciting cause, and I cannot accept the theory that it, and it alone, can cause chorea.

If we do, we must accept the fact, that all children are fortunate in not being daily frightened into choreic attacks.

I can understand how a child, with choreic dyscrasia ready to bud and blossom, may be incited to an attack by any reflex cause. If we accept fright, why not difficult dentition, phymosis and motor-ocular irregularities? They all certainly bear an equal relationship.

There is hardly a case of chorea in boys in which we do not obtain some degree of phymosis and a history of shock. It is a clinical fact, motor-ocular irregularities are common, also, and may be conjoined with the others.

A constant neuro-vascular excitement, extending over a period of time, would tend to develop an attack more surely than sudden severe impressions.

After we have chorea developed, slight irregular variations from any normal sensory motor or vaso-motor excitement may intensify or light it up again. This is logical, I think.

What complicates the adjustment of shock to chorea, is the refusal to recognize a pre-existing defect—a pre-existing morbid element at work.

How much easier to understand if faith is reinforced?

How pretty is this lesson in the case I have related? There was a most intense shock, but chorea was not developed till over six weeks afterward.

If possible, let us all admit this fact: that chorea is an expression of something else than a mere rampant neuro-motor mechanism due to shock or any other reflex cause.

Arguments are advanced that because we remove our irritating lesion chorea ceases. This is old, and only true in a very small minority of cases. I have seen chorea cease abruptly of its own accord. I have seen it cease from shock.

This to the clinician far from proves that chorea is only a symptom. The rarity of the abrupt termination of the neuro-motor phenomena is in itself a proof of a constant stimuli, acting by its ever being present till its impress or thralldom is at an end.

I would like here to state, that in the last three years I have not been able to satisfy my mind that I have seen one case which I could designate as a true reflex chorea. That is. one that depends upon a local peripheral irritation.

I can understand how, perhaps, a chorea might cause certain peripheral motor incoördinations, but I am not satisfied that oculo-motor troubles could be a prominent cause.

I, therefore, can not relegate a large percentage of choreas to a purely functional class, largely reflex and fostered by circumstances and tending to produce general functional disturbances.

To do so would, to me, be either admitted ignorance or biasness. A biasness which is discreditable to specialism in medicine and surgery.

Dispense with such a theory, and our little patients will receive a more needed rest.

Further experimentation in the field of reflex surgery ought to be abandoned.

Christian A. Herter has drawn attention to the fact that excessive excretion of uric acid was a highly con-

stant feature in pronounced chorea. In many cases of chorea, the urine had a reddish hue. There was a diminution of the chlorides in relation to urea. Almost all were of a high specific gravity (1024-1030) and of small volume in the twenty-four hours.

Work along this line of investigation will in future be of great importance. Cases in which I have been able to study the urine in reference to spinal gravity, they were all above normal. None, however, exceeded 1025, and all acid in reaction.

The most common nervous affection of rheumatism is chorea.

In our case reported we have rheumatism, not only well marked, but in conjunction with other correlative complications, observed the most frequently in childhood—growing pains, articular troubles, anæmia, cardiac lesions and the subcutaneous nodules.

The rheumatic diathesis in children is not to be expressed in the same way as in the adult. The latter shows a more frequent manifestation in muscles and joints, tenderness and swelling, fever, sweating, etc. In childhood it may only be expressed in the so-called growing pains, erythema nodosa, slight stiffness, etc.

Rheumatism is essentially a disease of the motor apparatus. Chorea is a disease of the motor centres. The analogy is interesting. While we recognize that rheumatism often immediately precedes an attack of chorea, there are but few cases observed where the onset of the chorea is immediately followed by an attack of rheumatism.

In the Guy Hospital Reports for 1890, an analysis is given of 262 cases of chorea from 1879 to 1889.

In not a single case was rheumatism seen in less than a week of the commencement of the chorea attack.

Thirty-four cases the rheumatic phenomena occurred between a week to a month; in sixteen cases it preceded the attack of other rheumatic phenomena by months or years.

I have noticed many cases in which rheumatic phe-

nomena, such as articular swellings, exudations and cardiac complications, did not make their appearance till months and even years after the chorea.

Not one case can I remember where it immediately supervened on the initial choreic phenomena.

Is it not logical to suppose many choreics, which are classified statistically, as non-rheumatic, which, if followed up, would be of this predicatory class? Statistics of chorea should be based upon cases observed for a long time.

As we often see other rheumatic phenomena alone in children, so can chorea be the only objective symptom emphasized. As we have seen in the predicatory cases, chorea seems to hold control of the other complications. It monopolizes, as it were, the attention of the rheumatic irritant for a more or less length of time.

A most instructive and interesting case appears in the Guy's Hospital reports, quoted above. A chorea of three years standing in a case, thirty-seven years of age, which had run the gamut of all the London hospitals with no apparent indicative complicating symptoms or previous history as a clue. He was only eleven days in Guys, when a severe acute attack of rheumatism appeared with accompanying cardiac troubles.

Two relapses occurred. He eventually left the hospital with improved heart and chorea. I have had under my observation, the past eighteen months, a similar case—a man of thirty-eight—with eudocarditis. Apical murmur and heard under the scapulas; muscular and articular pains in extremities with just a noticeable chorea face, tongue and extremities, and a history of a slight chorea at eleven years of age.

Of all the cases studied at Guys, there were seven only beyond the age of puberty. Those which were not associated with rheumatism, either received injuries or it occurred in young women at childbirth.

*Pathology of Rheumatism in Children.*—The articular manifestations are rather a vascular than a parenchymatous process, with little serous exudation. Tissue

elements are unaltered, and solid products or pus are rare : reabsorption is complete, and no traces are left.

Visceral complications suggest similar reflections. As Fernet says, touching without profoundly modifying, rarely leading to organic changes.

An intermediate between congestion and a true inflammation.

There appears often a more or less overgrowth of the fibrous tissue elements of an affected region.

Rise of temperature is little marked.

Chorea is rarely associated with rheumatism in the adult, and rarer yet the subcutaneous nodules.

It is an admitted fact by English observers, that the subcutaneous nodule is *always* accompanied with rheumatism, and no other disease. If it is present it is *absolutely* associated with other rheumatic phenomena, either in articular swellings, cardiac lesions or chorea. In the majority of cases on record there is associated all three correlative phenomena.

They were first noted by Hillier in a case of chorea, in 1846. Meynert, Barlow and Warner, Rehn, Hirschsprung and Cheadle all emphasize their rheumatic origin and common association with chorea.

Cheadle reports the fact of cases in which chorea and the subcutaneous nodules were associated alone. G. Wallace Anderson, *Glasgow Medical*, 1891, reports a case of nodules in a woman of fifty. Angel Money, *Lancet*, 1891, reports a case where they were found under the pericardium on the heart in a case of pericarditis.

Pathologically the subcutaneous nodule is an exudate and connective tissue hyperplasia with abundant vascular supply. The connective tissue is made up of loosely woven, wavy bands. They break down easily and by resolution disappear more or less quickly.

In the subcutaneous nodule we have the key to a homologous exudate and connective tissue hyperplasia that composes the little fibrinous nodules on the valves, and also the little colloid and hyalin bodies found in the choreic brain.

These hyalin exudates have been noted by Meynert, Dickinson, Berkely, Jakewenko and Dana.

The problems relative to the nature of humoral pathology are still unsolved. Whether there is a microbic factor as a starting point is not certain.

Until we can solve the nature of rheumatism, that of chorea will be mysterious.

Blake writes of a supræmic origin of rheumatism.

Distemper causes chorea in dogs. Distemper is allied to scarlet fever, the chorea is a late complication.

Sturges says: Chorea and rheumatism are, it is probable, members of a pathological group, having arthritis for a common factor, and of whose underlying source we are yet in search.

Chorea has also lately been transmitted by inoculation from one dog to another.

While I do not claim that the rheumatic poison is always the cause of chorea any more, as Cheadle says, than it is the cause of all joint inflammations.

Yet, as it is the most common cause of joint affections and is always the cause of the subcutaneous nodules and as chorea is so intimately associated with these nodules a relation much more commonly exists than is generally accorded to rheumatism.

Lastly, the motor centres stand in anatomical relation with rheumatism, as all other motor apparatus and fibrous tissue generally.

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