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A CLINICAL RE-EXAMINATION OF THE MOTOR-SYMPOMS OF CHOREA.¹

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AND

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of Philadelphia.

IN 1881 one of the authors of this essay pointed out that the previous descriptions of Sydenham's chorea were imperfect, and that its symptoms admitted of classification into clinical species or at least varieties. Taking Dr. Weir Mitchell's² grouping as a basis, the cases at his clinic have within a year been re-examined with care and with corroborative results.

From the text-books one is led to believe that the movements of chorea follow a definite type in all cases, with only occasional variations. It is claimed that the movements are continuous when the disease is advanced, rendering voluntary muscular acts impossible or capable of completion with the greatest difficulty; that the movements are increased by voluntary acts, though sometimes for a time quieted during such effort. The movements are said to be at first simply awkward and incoördinate—a condition which gradually develops into a constant, irregular spasm, clonic in character. This includes practically all that can be found in the books relating to the movements of chorea, save the delineation to be found in Dr. Weir Mitchell's lectures, to be quoted later.

In this study an attempt has been made to elaborate

¹ Read before the College of Physicians of Philadelphia, May 5, 1897.

² Lectures on Nervous Diseases of Women. By Dr. S. Weir Mitchell, 1881.

more fully the descriptions of the choreiform movements. Some of the cases cited present symptoms familiar enough, and are examples of the type usually described. They are briefly reported in order to complete the picture of chorea and that they may be contrasted with the less familiar and, if not unrecognized, certainly undescribed types. The histories are purposely condensed, since much of the data does not in any way pertain to the subject under discussion, but full notes of all the cases are accessible.

CASES ILLUSTRATING THE FIRST TYPE OF MOVEMENTS:

CASE I.—F., aged 10, the eleventh attack. The movements were general, worse on the right side, and continuous during rest. When two points were brought together the movements ceased or lessened greatly. The coat was buttoned without difficulty and a glass of water put to the mouth without the slightest danger of spilling. It may be truly stated that during fine, voluntary effort there was a total absence of the jerking.

CASE II.—F., aged 6½, the first attack. The movements were continuous and distinctly choreiform, and there was at the same time a suggestion of tremor. There were alternate flexion and extension of the hand 75 times a minute. Movements were entirely absent when she attempted to put a glass of water to her mouth, while writing her name, and when sewing. She also was able to pick up a spoonful of water without spilling it—in other words, small, fine, muscular acts occasioned an entire disappearance of the movements for the time. When the muscles again resumed a position of rest, at once the choreiform movements recurred.

CASE III.—M., aged 10, the first attack. The movements were severe, general, and continuous. Two points were brought together quietly, a glass of water was held at arm's length without spilling, and all signs of the movements disappeared while he wrote his name. When a complicated act was attempted, like buttoning the coat, there was for the time an increase.

CASE IV.—M., aged 8. The movements were continuous and general when at rest. They ceased entirely upon every attempt at voluntary muscular effort.

CASE V.—F., aged 10, the first attack. There were continuous general movements. Two points were approximated well, and a glass of water was carried to the mouth quietly.

CASE VI.—M., aged 12, the first attack. Movements were continuous and general, disappearing entirely when endeavor-

ing to bring two points together, or when the other tests were employed.

CASE VII.—M., aged 7, the first attack. There were general continuous movements, worse on the right side. They disappeared when writing his name as well as when other voluntary acts were made.

CASE VIII.—F., aged 10, the first attack. There were continuous general movements, quieted by intentional efforts.

CASE IX.—M., aged 8, the first attack. There were general continuous movements. All the movements disappeared on fine muscular acts.

CASE X.—F., aged 16, the first attack. There were continuous left-sided movements, which totally stopped when voluntary acts were attempted. She drew with surprising accuracy a straight line, free hand. Two points were brought together perfectly, with no sign of unsteadiness during the act.

CASE XI.—F., aged 8, the first attack. The movements were right-sided and continuous during rest. In making an effort to write her name the movements for the time disappeared entirely, and a glass of water was carried to the mouth with perfect muscular composure. This was the invariable result of voluntary muscular effort in this case.

CASE XII.—F., aged 13, the second attack. There were right-sided continuous movements. She wrote her name quietly, and other muscular efforts effected for the time total disappearance of the movements.

CASE XIII.—F., aged 14, the first attack. There were general continuous movements. Two points were brought together quietly; raising a glass of water to the mouth quieted the movements—in fact, all muscular effort short of fatigue caused the movements to cease for the time.

CASE XIV.—M., aged 11, the fourth attack. There were continuous left-sided movements. Two points were quietly approximated and a glass of water held without spilling. Any effort causing fatigue or needing continuous exertion increased the movements.

CASE XV.—F., aged 20, the first attack. The movements were left-sided and continuous. This was a striking case of pure, passive chorea. Upon any muscular effort of the involved muscles the choreiform movements vanished entirely. The patient was able to draw a straight line, to cut faultlessly with a pair of scissors along a line drawn on paper, and to drink from a glass held in the hand, without the slightest evidence of the disease.

CASE XVI.—M., aged $7\frac{3}{4}$, the first attack. General continuous movements were quieted by volition and fine muscular efforts. Two points were approximated perfectly, every sign of chorea for the minute disappearing. (*See writing.*)

Willie P
Hacker

CASE XVII.—F., aged 6, the first attack. The movements were continuous and general. When attempting to hold a glass of water in the hand or to perform delicate muscular acts the movements disappeared. Violent muscular action increased them everywhere; fixed attention had no effect. (*See writing.*)

Katie Simon

CASE XVIII.—F., aged 13, the third attack. There were marked, continuous movements. She held a glass of water at arm's length motionless, brought two points together quietly, and wrote her name without the slightest obvious trouble.

CASES ILLUSTRATING THE SECOND TYPE OF MOVEMENTS:

CASE XIX.—M., aged 12, the first attack. Movements were general, worse on right side, continuous during inaction. The movements when at rest were moderately severe, but the slightest attempt to use the hands increased them markedly. On reaching for a glass of water they became so immoderate as to render the completion of the act impossible. To write her name was quite impossible. She was incapable of buttoning her coat or otherwise dressing herself. While at rest the movements subsided largely, but never disappeared.

CASE XX.—F., aged 22. The movements were continuous and general. A glass of water when held out from the body was spilled, and other voluntary muscular effort increased the movements.

CASE XXI.—F., aged $7\frac{1}{2}$, the first attack. There were right-sided continuous movements. Fine muscular effort increased them. (*See writing.*)



CASE XXII.—F., aged 10, the second attack. There were general continuous movements, worse on the right side. She spilled a glass of water when raised to the lips. Movements were increased on trying to write, to bring two points together, or to make any fine muscular effort.

CASE XXIII.—F., aged 4½, the first attack. The movements were general and continuous, worse on the right side. Fine volitional effort and also violent coarse muscular efforts increased them.

CASE XXIV.—F., aged 19, the second attack. There were general, continuous movements, increased when attempting to put a glass of water to the mouth or to bring two points together.

CASE XXV.—F., aged 6, the first attack. There were left-sided continuous movements. A glass of water was spilled when held in the hand. Fine muscular acts and efforts of volition increased the movements. When attempting to fasten her shawl with a safety-pin, the incoördination became so excessive as to make the act quite impossible.

CASES ILLUSTRATING THE THIRD TYPE OF MOVEMENTS:

CASE XXVI.—M., aged 5, the first attack. The movements were confined to the right side, and during rest were practically absent, only an occasional slight jerk being observed from time to time. When an attempt at voluntary movement was made, such as raising a glass of water to the mouth, the twitching became at once so violent as to spill the water. An attempt to write his name induced extremely incoördinate movements. The choreiform motions were practically absent until muscular effort was made.

CASE XXVII.—F., aged 13, the first attack. Movements during rest were absent entirely for varying periods, or, if present, only of slight degree. The smallest muscular efforts, however, threw the group of muscles used into the most surprising condition. Violent large choreiform movements appeared and continued while the effort lasted. Afterward they disappeared almost entirely, and only rarely recurred during inaction.

CASES ILLUSTRATING THE FOURTH TYPE OF MOVEMENTS:

CASE XXVIII.—F., aged 11, the first attack. The movements were rhythmic and rotary, yet possessed the character of choreiform movements. They were not continuous. The various tests used failed to make any per-

ceptible difference in the character of the movements. When a glass of water was placed to the mouth, the name written, or any fine or coarse muscular effort attempted, the movements persisted, unaltered in kind and extent.

CASE XXIX.—F., aged 6, the second attack. There were continuous and general movements. Here again voluntary muscular acts, fixed attention or effort of the will altered the movements little or not at all.

CASE ILLUSTRATING ADMIXTURE OF TYPES:

CASE XXX.—F., aged 9, the second attack. There were right-sided continuous movements, which finally became general. She wrote her name with perfect ease, the movements ceasing entirely. Other fine muscular acts gave similar results. A second examination, made about a fortnight later, disclosed a changed condition. The movements during rest were barely perceptible, but became marked when she attempted to button her coat or to make any other similar muscular effort. It was learned that the same condition had probably existed at the onset of the disease. Within a short time this case presented symptoms of two distinct types: First, the condition in which voluntary muscular effort was necessary to render incoördination manifest; secondly, the state in which movements were continuous and increased by voluntary effort.

These cases, all examples of Sydenham's chorea, present, in that aspect of the clinical history which relates to the movements, a striking diversity. Four different types of movements may be distinguished. In the first type the movements during periods of muscular inaction are continuous. The hands lying in the lap of the patient are in constant motion. But when the patient raises a glass of water to his mouth or performs any act requiring a like delicate muscular effort there is witnessed an entire disappearance of the movements, lasting throughout the muscular effort. This appears to be a phenomenon quite beyond the influence of the will, and suggests that inhibition, for the time, is increased. (*Cases I to XIII inclusive.*) In a second type there is a continuous clonic contraction of the affected muscles during rest, while every attempt to perform muscular acts is attended with a surprising increase in the violence of the jerking, apparently independent of

and beyond volitional control. It is impossible for one suffering from this variety of the malady to complete satisfactorily any voluntary muscular act. This condition suggests, contrary to that presented by the first type of movements, a lessened inhibition manifested during willed muscular efforts. This is the usual type described. (*Cases XIV to XXVI inclusive.*) A third, and the rarest type of the movements, is that which becomes evident only on attempts to perform a muscular act. The hands at rest move, if at all, only slightly and at rare intervals; but on attempting to use them the twitchings become sufficiently active to prevent or greatly to interfere with the performance of the act. (*Cases XXVI and XXVII.*) A fourth type is illustrated in a small number of cases, in which the movements, continuous during rest, are but slightly altered by the tests employed. (*Cases XXVIII and XXIX.*)

It is the third type which presents points of unusual interest. It might be claimed that the cases illustrative of this type were only in an incipient stage of development, when such a condition might be expected; but in the two cases cited (*Cases XXVI and XXVII*) the movements which took place upon applying the tests were so marked as to clearly demonstrate that the disease was fully developed. Furthermore, in one case (*XXVI*) the same type remained unchanged throughout the whole course of the disease. In one case (*Case XXX*) two types were observed, appearing at different stages of the disease. Voluntary effort was at one time the signal for an entire disappearance of the movements. There was, however, a history of at times spilling water from a glass and dropping things from the hands. It is difficult to say to what extent this admixture of the types existed in these children, as many of them were out-patients, and they could be seen but once a week. It seems to us, however, that in

some cases the same type may persist throughout the disease, and that, on the other hand, more than one type may be present in the same case at different stages of the disease, though this requires further study.

It appears, then, that there are: 1. Cases of chorea which show, some at one stage of the disease, some throughout their course, an absence of movement during rest, requiring muscular action to develop what may be either mild or severe choreiform movements. 2. There are case in which the movements are continuous during rest, but become greatly increased on intentional effort. 3. There are cases with severe choreiform movements which disappear entirely when muscular acts are performed. 4. In some cases the movements seem to be unaltered by voluntary muscular efforts. 5. There are cases which present during their course at different times more than one of the types described.

If Sydenham was familiar with these phases of the affection, we can find no evidence of it in his descriptions. He states:

“If the hand of the same side be applied to the breast or any other part of the body, the child cannot keep it a moment in the same posture, but it will be drawn into a different one by a convulsion, notwithstanding all its efforts to the contrary.

“Before a child who hath this disorder can get a glass or cup to his mouth he useth an abundance of odd gestures; for he does not bring it in a straight line thereto, but his hand, being drawn sideways by the spasm, he moves it backward and forward, till at length the glass accidentally comes nearer his lips, he throws the liquor hastily into his mouth and swallows it greedily, as if he meant to divert the spectators.”

We can find no careful description of these varieties of choreiform movements earlier than that in Dr. S. Weir Mitchell's *Lectures on Nervous Diseases*. In his chapter on chorea he makes the following classification: 1. Those in which there are awkwardness and

incoördination of voluntary movements, followed sooner or later by automatic or unwilled clonic spasms of various parts; 2. Irregular movements, only occurring during willed actions; 3. Constant automatic, irregular clonic spasms, usually of the hands; but during volitional acts these entirely vanish, and the most complicated acts are well performed without obvious incoördination.

Bernt (1810), quoted in Osler's³ monograph, recognized that in some cases the movements disappear when the patient stands up. Osler quotes also Dr. Weir Mitchell's classifications without further comment. Robertson, in 1805, stated that the patients affected with this disease have no control over their actions by any effort of volition. In his discussion of the movements of chorea Gowers asserts as typical of the disease that the movements are continuous and violent, and that they are "always increased by excitement and by attempts at voluntary movement;" and, further, "voluntary movement is always, in severe cases, irregular;" but, on the contrary, we have seen and cite in this paper many cases in which extremely violent movements vanish entirely during the voluntary act. Dertz, in an inaugural dissertation on chorea, published in 1872, refers to the increase of the symptoms when a voluntary effort is made, and states that volition quiets for the time, only to be followed by an increased amount of motion. Sturgis,⁴ in 1893, described two varieties, occurring in young children, of long duration and of moderate degree: 1. The symptoms were best when the patient was unnoticed; 2. Efforts of will showed a considerable power of control, and simple muscular acts without the control made apparent the unsteadiness of the disorder. Ziemssen (in

³ Osler: Chorea, 1894.

⁴ Sturgis: Chorea. London, 1893.

Sturgis) considers the incessant twitchings as characteristic, while they may be sometimes spontaneous and sometimes excited by voluntary impulses. More recently some have recognized, though not to its full importance, that on voluntary muscular action the movements may disappear (Blocq⁵ and Sinkler⁶). Similar quotations could be multiplied indefinitely, with a fuller or more accurate description of this phase of the disease in none.

What significance these facts possess we shall not attempt to say, but they are certainly of undoubted clinical interest.

There can be little doubt that Sydenham's chorea is a disorder the seat of which is in the motor areas of the cerebrum. If there were any doubt as to this it should be settled by the cases in which distinct local trouble of these regions, specific or from a thrombosis, has given rise to typical unilateral choreoid motion. In the various forms of post-hemiplegic disturbance of motion may be observed in imperfect shapes all the types of motor-disorders set forth in this paper.

Chorea is, however, a very distinct clinical grouping of symptoms; and it may be better, on the whole, to reserve for it the label chorea, and so to speak of the post-paralytic motor-symptoms as choreoid or choreiform. Locally, they offer unmistakable resemblance to the malady chorea; but they also may present, when serious, mental and other peculiarities not to be met with in the choreoid products of local disease of the motor-areas and perhaps of the internal capsule. A more complete study of these is to be desired.

⁵ Blocq : *Traité de Médecine*, Paris, 1894.

⁶ Sinkler : *Chorea, A Text-book on Nervous Diseases*. Edited by Dercum, 1895.

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