AN ANALYSIS OF SIXTEEN CASES OF CHOREA AND MOTOR TIC.

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In the present paper the term “chorea” is used to connote the presence of sudden inco-ordinated and involuntary movements of a purposeless character, partial or generalised in their distribution, and which cease during sleep. Although the majority of the cases quoted belong to the classical type of Sydenham, others, whilst presenting an identical group of symptoms, differ from them chiefly in their chronicity. Similarly, the cases of motor tic are included because there are sound reasons for considering them in this connection. It is a matter of common observation, for instance, that a motor tic affecting single muscles or groups of muscles is not uncommonly left as a vestigial affection after an attack of chorea. Case III. (M. R.) is an instance of this. Moreover, the muscular contractions occurring in chorea are found to vary in intensity within the widest limits, ranging from the merest twitching of individual muscles (as in Case II., A. H.) to the most violent jactitations of the whole body (Case XII.), so that a case of typical chorea often passes insensibly into one of motor tic. The fact, too, that motor disorders, whether choreiform, athetoid, or rhythmical, are so frequently found in conjunction, and on an identical pathological basis, constitutes an à priori ground for considering them together.

As to the type of material dealt with in this paper, it is necessary to state that 13 of the series occurred either in association with amentia or in near relatives of aments, the other 3 being of the class met with in ordinary practice. The former group comprised the total number encountered during the routine examination of 80 consecutive cases of amentia of all degrees. It is important to bear this in mind when we come later to the question of etiology. Of the total number of cases described only a comparatively small number came under actual observation during the attack, but were elicited during an inquiry into the family history. Of the cases so obtained details are necessarily meagre, but were sufficient to satisfy one as to the nature of the complaint. Finally, of the 16 patients described, 10 were examples of acute chorea, 2 of chronic chorea, and 4 of motor tic.

The following is a brief summary of the cases:—
(A) Acute Chorea.

Case I.—E. S., age of onset 17. Duration of illness about 4½ months.

Family History.—Was one of 6 children; father was an epileptic, mother died of pulmonary tuberculosis.

Medical History.—No history of rheumatism or previous attacks of chorea.

Condition When First Seen.—A thin, anæmic girl. She showed constant choreiform movements of left arm and leg, and twitching and grimacing of the left side of the face. She had to hold her left arm forcibly to restrain the movements—could not hold things with it, or feed herself. The left foot was dragged in walking, and the disorder was markedly increased during voluntary movements. The symptoms ceased during sleep and were increased during observation. There was no change in the affective mental condition, except a slight irritability. Examination of the heart revealed a soft systolic bruit (probably hæmic) heard in all the cardiac areas, and not conducted into the axilla; there was no temperature, and no arthritic or other manifestations of rheumatism. The symptoms set in about ten days after a severe fright. Going downstairs during the night she saw a moving object which she imagined to be a burglar, but which turned out to be only a cat. She was much startled by the occurrence, but apart from the mental shock appeared to be none the worse for some time. Subsequently movements began in the arm, spread to the leg, and lastly to the face.

Course and Treatment.—First seen six weeks after onset. Treatment was carried out by hypnotic suggestion. Later, she was put on a course of Blaud's pills. It was noticed during the first sitting that the movements were much less during hypnosis, but returned in full force immediately after waking. After a few sittings the movements entirely ceased during the actual "sleep." She was seen altogether about eight times, at intervals of about a week, at the end of which period she was sufficiently well to return to work. Improvement was first noticed in the face muscles, later in the arm, and last of all in the leg. At one period in the treatment there was a slight involvement of the right arm, which, however, never came to anything. The exciting cause in this case was undoubtedly the psychic trauma acting on an unstable nervous system. The girl had previously been in excellent health, and the anæmia was probably secondary to the chorea. Moreover, she was well on the road to recovery before the administration of iron was commenced.

I have dealt with this case at some length as it illustrates the part played by inherent neuronic instability, and is an
instance of chorea being set up by mental trauma and yielding to mental therapy.

CASE II.—A. H., age of onset 7½ years. Duration of acute symptoms about one month, though movements were still present two months after onset.

Family History.—He is one of two children. The other died after burns. Cousin on father's side is a mental defective and attends a special school. Another cousin in the same family has pulmonary tuberculosis. Father somewhat addicted to alcohol.

Description and Course. — When first seen had severe pain and swelling of knee- and finger-joints with fever. In addition, there was a systolic bruit heard in all cardiac areas, but loudest in mitral area, and not propagated into axilla. There was also a short diastolic bruit heard in mitral area. No displacement of apex beat or increase in area of cardiac dulness. Patient was very anaemic, and choreic twitching of mouth, lips, head, and fingers was noticed. Isolated muscles or groups of muscles appeared to be affected, especially the corrugator supercili, occipito-frontalis, and levator and depressor anguli oris in the face. In the hand the lumbricales and interossei were involved, and all the movements were increased during speech or on attempting to move. All superficial reflexes (including cremasteric and plantar) were much increased. Salicylates were administered and the acute symptoms subsided. The intensity of the movements varied directly with the temperature, but persisted to some extent after all acute symptoms had disappeared, as did also the cardiac bruit. It should be mentioned that the choreiform movements preceded the onset of the rheumatism by several weeks, and were then thought to be due to habit. At the present time, 3 months after onset, patient has a healthy complexion, and the twitching of the muscles of expression is barely discernible.

CASE III.—M. R., age of onset 7 years. Duration of attack about 10 months. Movements began in hands and spread to legs and involved both sides of the body. Confined to bed for two weeks. Present time (3 years after attack) he still has a slight twitching of the orbicularis palpebrarum, and marked paresis of upper extremities, both of which are said to date from attack of chorea. The paresis of the hands is so marked as seriously to interfere with his ability to write or do work requiring much manipulative skill. He is anaemic, but shows no signs of valvular disease of the heart. Has recently had two epileptic seizures—one a typical major attack with loss of consciousness, and the other Jacksonian in type.

Family History.—Only child. No neuropathic ancestry. Mother's sister died of pulmonary tuberculosis. Mother's mother has rheumatism.
Cases IV. and V. were both examples of acute chorea occurring in the relatives of an ament. The relationship in this case was that of cousin, and the conjunction of amentia, chorea, and tuberculosis in the genealogy appended is particularly to be noted.

Case VI.—F. N., at. 19. Had acute chorea at the age of 7 years, lasting 7 months. He is a feeble-minded youth and suffers from epilepsy. Fits began at 11½ years of age after a fall. Said to have been mentally bright before that. Now very much demented. Heart normal.

Family History.—Paternal grandfather was "paralysed" for 30 years. Mother and maternal grandmother had rheumatism, and the latter "died of dropsy." Maternal aunt is in an asylum. A brother of the patient is addicted to alcohol. A sister is delicate and "tuberculous," and another brother did not talk until the age of 3.

Case VII.—Acute chorea occurring at age of 13, after two attacks of acute rheumatism at the ages of 9 and 11 respectively. The affection lasted 9 months, and at present she has mitral disease. This girl has 3 sisters who are mentally defective, as shown in table appended.

It is to be noted that the chorea occurred in a member of the family who showed no mental defect, but in whom the neurotic predisposition was probably latent.
Case VIII.—M. R. Acute chorea in a mental defective, æt. 15. Had chorea at age of 12, and was at home from school nearly a year. Said to have had a "rheumatic heart."

Present Condition.—A feeble-minded girl with small cranium and low bulging forehead. Exhibits erotic tendencies. Is myopic and has right internal strabismus. Reflexes (superficial and deep) very sluggish. Heart sounds normal.

Family History. — Father was charged with attempted suicide, otherwise no neurosis.

Case IX.—I. C., acute chorea in an epileptic imbecile, æt. 17. No information as to attack.

Present Condition.—Has both major and minor fits. The former are infrequent and affect chiefly the right side. Has marked paresis of right leg below the knee, and drags right foot. Neurotic heredity shown in accompanying table.

Case X.—P. B., acute chorea in the sister of an epileptic idiot. No particulars. The child herself is almost completely paralysed in the lower extremities. Cannot stand or walk, or sit erect. She is unable to do anything for herself, and her mental status is that of an infant.

(B) Chronic Chorea.

Case XI.—A. E. S., æt. 13. Chronic chorea of 10 years' duration occurring in an imbecile. At the age of 2½ years she had a fall and injured her head. Was unconscious for a short time; subsequently developed a squint, and later chorea. Since then has deteriorated mentally.

Present Condition.—Exhibits constant choreic movements affecting the whole of the body. It is to be noted that the chorea only shows in the lower extremities when patient is sitting. There is some paresis of upper extremities, and grasp of hands is weak. The movements are extremely rapid, as is also her speech. In her conversation there is a certain resemblance to the "flight of ideas" seen in a case of mania, except that the flow of words is not so rapid. She talks incessantly—a good deal of it is nonsense—but some of her remarks are pertinent and relate to other people's conversation, here again resembling mania. Her speech is very imperfect and difficult to under-
stand and almost as "choreic" as her movements. Though apparently inattentive, her conversation is automatically directed by whatever strikes her attention. She is very small for her age—no doubt owing to her constant restlessness—but shows no evidence of disease apart from that described.

**Family History.**—Maternal cousin died from spinal affection, nature unknown. Otherwise no neurosis, no stigmata of degeneration, and physiological development proceeded on normal lines.

In this case both the amentia and the choreic affection were secondary to the fall in infancy. What the actual lesion may have been it is impossible to surmise, but in view of the mental defect and the generalised distribution of the chorea, it is highly probable that an inflammatory or vascular lesion of the cortex occurring at the time of the injury, acting as a permanent irritant, has induced an instability of the cortical motor neurones on the one hand, and an involution of the cells of the prefrontal region on the other.

**Case XII.**—H. B., æt. 14. "Chorea spastica," dating from birth, and involving the greater part of the body.

**History.**—This was an example of birth palsy (spastic diplegia). Patient was an eight months' child. Parturition was difficult, and child was said to be blue for some days after birth. Head is still much deformed; the moulding of the cranium is permanent and like that seen in an unreduced brow-presentation, viz. a high, precipitous forehead, sloping back sharply to the occiput. Physiological development delayed—has never walked and did not talk until 3½ years of age.

**Present Condition.**—He is grossly deformed, very big for his age, and well developed physically. The left leg is contracted, left arm and hand much contracted, and finger-joints hyperextended. Right arm still more deformed, and fingers are bent into the palm of the hand. Sight and hearing normal. Pupils equal; react to light and accommodation. He sits habitually with the left leg thrown over the right and the right hand held between the legs to restrain the violence of the choreic movements, which affect the left leg and right arm the most. The movements are violent, and involve the head and the whole of the left side, and, to a certain extent, the right arm. The contractions of the limbs are evidently spastic in nature, and any attempt to separate them or extend the joints is attended by a marked exaggeration of the choreic movements, so extreme as to throw him out of his chair. He cannot pick up or hold anything with his hands, of course, and cannot stand alone. He habitually uses his tongue to turn over the leaves of a book, and can seize and swallow his food off
the ground very much as a dog does. By dint of this extraordinary use of the tongue it is greatly hypertrophied and wrinkled (macroglossia). Both superficial and deep (tendon) reflexes are absent. His education has of course suffered considerably, though his mental condition is very fair considering his opportunities. Speech is staccato and explosive, but quite intelligible. Though described by the Education Authorities as mentally enfeebled, the defect is largely secondary to the physical disability and lack of education.

(C.) Motor Tic.

Case XIII.—G. D. M., æt. 9. Motor tic occurring in an imbecile (secondary ament).

History.—Affection dates from infancy. At 1¼ years of age she had pneumonia and meningitis. Subsequently there was paralysis of the left side, from which she has only partially recovered. In addition to the weakness of the left side the child was noticed to be dull mentally, and has never made headway. Movements commenced later in the left arm, and have continued up to the present time. No neurosis in family, and no stigmata of degeneracy.

Present Condition.—Has marked paresis of the left arm and leg, and walks with a decided limp, pressing on right side. Has constant rhythmic movements of the left arm and hand, in which she alternately grips the right hand and then relaxes her hold. Occasionally when her attention is excited the movements cease, and also during sleep. Has left internal strabismus and slight scoliosis. Patellar reflex present—increased on left side. Also has frequent attacks in which she falls to the ground, and which resemble epilepsy of the Jacksonian type. Mentally she is an imbecile of a low grade, cannot talk or feed or dress herself, and is quite incapable of education. The amentia, hemiplegia, and motor tic are all sequential to the attack of meningitis occurring in infancy.

Case XIV.—C. E., æt. 9. Motor tic occurring in an idiot (primary ament).

Family History.—A sister had “paralysis” at 13½, and suffered from some form of tonic spasms of the left arm—both of them probably hysterical in nature. A brother died in a fit at 2½ years. The mother stammers occasionally, and has a motor tic affecting eyelids on both sides. Physiological development much delayed.

Present Condition.—He exhibits constant clonic contractions of orbicularis palpebrarum exactly resembling that seen in mother, but said to have developed recently. Also has left internal strabismus and some photophobia.

Mental Condition.—Is continually restless—utters peculiar and stereotyped cries. Interferes with and touches everything in neighbourhood. Cannot speak or do anything for himself. Resists examina-
tion. Is destructive, noisy, and boisterous, and interested in nothing. Mentally he is an infant.

Case XV.—F. M. H., æt. 15. Athetoid movements of left hand in a feeble-minded epileptic. Fits began immediately after birth, and are of both major and minor type, occurring on an average once a fortnight.

Present Condition.—Marked scoliosis; right hip much higher than left. Right genu valgum. The left arm and hand are paretic. Vision of left eye is weak. Speech normal. Mentally she is facile and easily amused, and undoubtedly feeble-minded.

Family History.—Patient is one of four children. The paternal grandfather was an alcoholic. Mother had five miscarriages (?syphilis).

Case XVI.—J. S., æt. 12. Motor tic in an epileptic imbecile (primary ament).

Family History.—Patient is one of ten children (8 living). The other two died in fits, one in infancy, the other at 9 years of age. Physiological development much delayed—walked at 5 years of age, did not talk until 8 years old. Fits began at age of 6 weeks, and have continued up to present time.

Present Condition.—Shows numerous scars due to falls during fits. Tongue is much furrowed. The right ear is malformed. Fits are of major and minor type. In addition he shows constant clonic and tonic movements of the head and eyelids, and at frequent intervals half closes his eyes. The clonic contractions rather resemble the intentional tremor of disseminated sclerosis, but are coarser in type and continuous. Mental status is low and bordering on idiocy. Some slight power of comprehension, but does not respond to questions. Cannot wash or dress himself, slavers, and is wet and dirty. He is obstinate and noisy, and exhibits echolalia to a pronounced degree.

From the study of the above cases certain facts emerge, which may be briefly summarised as follows:—The most predominant is the existence in the majority of the cases of a neuropathic basis for the affection, as evidenced by the co-existence of other neuroses or abnormalities of the nervous system, either in the individual case or in other members of the family. Seeing that the majority of the cases (13) were associated with amentia, or occurred in near relatives of aments, this is only to be expected. It is to be remarked, however, that of the 13 cases so described, 7 were examples of the acute (Sydenham’s) chorea met with in ordinary practice. Three cases which lie outside this group (Cases I., II., and III.) were also of the acute type, and were equally associated with neuroses, either in the same individual or in near relatives.
Of the 16 cases dealt with, 8 were associated with epilepsy; in 3 the patient alone was affected; in 2 it occurred also in a relative; and in 3 near relatives were affected. Here, again, the class of material dealt with must be remembered, epilepsy being so frequently associated with amentia. Aldren Turner, it may be remarked, states that epilepsy is found as an antecedent factor in chorea to the extent of 14·2 per cent.—the term being used here to denote chorea minor. He goes on to say: “It is obvious from a study of the literature of this subject that an intimate relation exists between epilepsy and chorea. The facts narrated show that chorea may predispose towards epilepsy and epilepsy towards chorea, the latter being the more common in my experience. In a neuropathic family epilepsy and chorea may be present in different members, e.g. one child may be an epileptic and another have chorea—an alcoholic father had two children, one an epileptic and the other choreic. Epilepsy and chorea may be present either simultaneously or at different times in the same person; in one such patient an attack of chorea was followed by freedom from fits for a year.” The co-existence of epilepsy and chorea in different members of the same family is strikingly illustrated in Cases VII., IX., and X. Case III. is an example of chorea followed by epilepsy later in life. Finally, of the 8 cases associated with epilepsy, three were instances of motor tic.

As to the part played by rheumatism, it is noteworthy that in only 4 of the series was there an undoubted history of rheumatism. In one (Case II.) the chorea was associated with a definite attack of acute rheumatism and endocarditis; in another (Case VII.) it had been preceded by two attacks of acute rheumatism, and there was permanent involvement of the mitral valves; whilst in the third (Case VIII.) the patient was said to have had a “rheumatic heart” at the same time as the chorea. In Case VI. mother and grandmother had suffered from “rheumatism,” and the latter had died of “dropsy.” As, however, no details were available in several cases, it is possible that the part played by rheumatism was greater than these figures indicate. All four cases mentioned were of the acute (Sydenham) type.

There is a definite history of tuberculosis in six members of the series, all of them being cases of acute chorea. In two of them (Cases IV. and V.) the patients were cousins, and the occurrence of tuberculosis, chorea, and amentia in different members of the family is strikingly illustrated (vide diagram). Here, again, the definitely established relationship between tuberculosis and
the neuropathic diathesis is partly to be held accountable, in view of the material dealt with.

Out of the total of 16 cases, 9 occurred in conjunction with some degree of amentia, and these included three examples of acute type, two of chronic chorea, and four of motor tic. Further, of the 6 examples of the chronic type of motor affection, the majority (4) occurred in patients the subject of secondary amentia, 2 only being found in primary aments. The former showed either in the history or clinical picture evidence of gross cerebral disease occurring in infancy, and including respectively meningitis, hemiplegia, and spastic diplegia (birth palsy); one only (Case XI.), though undoubtedly traumatic in origin, not revealing any indubitable evidence of a localised lesion. In this connection Tredgold, speaking of primary amentia, says: "Chorea is not common, but is found in some instances. Various forms of athetosis are fairly frequent in the severer grades. Intention tremor is occasionally seen. . . ." Later, in discussing the motor affections of secondary amentia, he says: "Jacksonian or epileptic convulsions occurred in a considerable proportion, whilst athetoid or choreiform movements are also frequently seen." Again, speaking of paralytic aments, he says: "In a few cases there is seen a constant rhythmic tremor or irregular choreiform movements without epilepsy," from which one gathers that chorea and allied conditions are, on the whole, more common in secondary than in primary amentia. Moreover, it is obvious that by chorea the chronic type of affection is meant.

Conclusions.—The limitations, both as to amount and type of material, here dealt with would preclude one coming to any dogmatic conclusions, but it may be helpful to see how far the findings agree with the accepted views as to the etiology of chorea. Reasons have already been adduced for grouping and considering these various affections under one head. It is of course usual to regard the acute chorea of children as a definite clinical entity in no way related to the chronic type of affection associated with organic disease of the cerebrum. Even when the term chorea is so delimited, there is no consensus of opinion as to its causation. Without going into any detail, it may be said that the general view is that it is merely one of the manifestations of acute rheumatism. Its frequent association with arthritic symptoms or with endocarditis, and its occurrence either in conjunction with rheumatism or as an alternative affection, have been urged in this connection. At the same time, many writers have
insisted upon its neuropathic relationships, notably Sturges (quoted by Osler 5), who regarded it as "an expression of the functional instability of the nerve centres." The frequent occurrence of neuroses either in the individual or family history, its occasional supervention after severe fright or shock or school-strain, the related nervous and psychical symptoms, and its preponderance in females are among the reasons which have been adduced in favour of a neurotic origin. Galabin, e.g. in discussing the chorea of pregnancy, says: "It cannot be doubted that pregnancy promotes the disease in two ways: first, as a cause of reflex irritation, and, secondly, by impoverishment of the blood. The element of mental emotion, well known as a starting-point of chorea, is also added in some cases, as when an unmarried girl becomes pregnant. Those who suffer from chorea in pregnancy are generally young primiparæ, who have either suffered from the disease as children or have an hereditary tendency to neurosis." It is possible that by a too rigid adherence to the accepted classification the underlying neurotic basis has not been apportioned its due value. At all events, it may lead to a broader conception of the underlying cause if one regards chorea merely as a syndrome, varying in intensity and in duration and in clinical setting, and seek to determine the constant factor, if such there be, common to all the conditions under which it occurs. As Ivy Mackenzie 7 rightly says, "if the phenomena are invariably the same, independent of the other pathological processes with which they are associated, there must be some common factor to account for the condition in all cases." He goes on to say, 8 "the point of view which supplies a conception of this common factor is that which regards chorea as a functional disorder of an unstable brain, such instability being due in some cases to constitutional causes, in others to toxic exhaustion, and in still others to organic brain disease." It may be helpful to examine how far this view of the case accords with the result of the present analysis. One may assume that all the examples of acute chorea described in the present series fulfil the requirements of the above-stated view, inasmuch as they all afford evidence of a neuropathic constitution, accompanied in some cases by a rheumatic toxæmia. Although, for reasons above stated, the neuropathic background is more evident than it would be in an equal number of "average" cases, it must be borne in mind that the cases cited are of the classical type met with in ordinary practice, and it is possible that a systematic inquiry into the individual and family history of every case of chorea would
reveal a larger proportion of neuropaths than is generally conceded. In any case, the comparatively large number of cases of chorea and motor tic (13 out of the series of 80), either directly associated with amentia or occurring in near relatives of aments, is self-significant. With regard to chronic chorea and the motor tics the case is rather different. Of the 9 cases directly associated with amentia, it will be noted that in 3 it took the form of acute chorea, in 2 the chronic form, and in 4 that of motor tic—in other words, a chronic affection was the more common. Moreover, in the six examples of chronic type the majority (4) were found in association with secondary amentia, that is, with definite cerebral lesions. These were various in type, and comprised infantile meningitis (Case XIII.), hemiplegia (Case XV.), Little’s disease (Case XII.), and one case (XI.) in which, although the movements were generalised, there was strong evidence of traumatic origin, resulting in an irritant lesion of the cortex. In other words, in most of these cases there is evidence of definite pathological change in the cells of the psychomotor cortex or their downward prolongations, and as the motor affection is found usually to be confined to the part of the body innervated by the damaged area, it seems reasonable to allocate to the latter an anatomical basis for the disorder, whatever type it may assume.

The two cases of motor tic in which there was no evidence of definite cerebral lesion, and which might appear to traverse this view of the case, were of such low mental status—one being an idiot and the other bordering on idiocy—as to suggest a very rudimentary development of the cortical neurones. Here one can hardly agree with Mackenzie, who regards all cases alike as “a functional disorder of the nervous system,” which, in the type of case just described, “has been rendered unstable by the organic brain disease.”

The evidence adduced would seem to assign to the neuropathic constitution a preponderating rôle in the etiology of chorea and allied disorders, but discloses strong reasons for differentiating, clinically and pathologically, between the acute type of short duration and the forms associated with states of cerebral subevolution and degeneration, whether these be of the primary neuronic type or sequential to vascular or inflammatory lesions.

References.—1 Aldren Turner, Epilepsy, p. 158. 2 Tredgold, Mental Deficiency. 3 Ibid. 4 Ibid. 5 Osler, The Principles and Practice of Medicine, 4th ed. p. 1082. 6 Galabin, A Manual of Midwifery, 6th ed. p. 350. 7 Ivy Mackenzie, “Chorea,” Glasgow Med. Journ., June 1915. 8 Ibid. 9 Ibid.