NOTES FROM A CLINICAL LECTURE ON A CASE OF HUNTINGTON'S CHOREA.

DELIVERED AT ADDENBROOKE'S HOSPITAL, CAMBRIDGE.

BY

PROFESSOR SIR CLIFFORD ALLBUTT, M.D., F.R.S.,
PRESIDENT OF THE BRITISH MEDICAL ASSOCIATION.

GENTLEMEN, I have asked this patient to present himself here this morning in order that you may see a case of a very infrequent disease; I cannot recall more than four such cases, if indeed so many, in my own experience. The title of Huntington's chorea I retain because Huntington—a physician of New York—published, in 1872,¹ the first comprehensive description of the malady. It seems to have had some endemic prevalence in Long Island, New York, as Huntington's father and grandfather and other physicians in the district had observed and distinguished it. The Huntingtons laid much stress on the hereditary incidence. This record fell into neglect until 1884, when Ewald recalled attention to it. Many cases have since been described in both hemispheres, some with necropsy.² The alternative title of Hereditary Chorea I avoid because in many instances it has not been obviously hereditary, and no such condition is present in the case now before you.

At one of our Discharged Soldiers' Employment Committee meetings the last case on the day's list was of a man, X. Y., who had been sent from one hospital to another as a case of "neurasthenia"; and ultimately discharged. I was thinking it not worth while to wait longer for one of this crowd when the door opened in an irresolute way, and an arm was thrust through with a spasmodic jerk; then a leg followed with a like unsteadiness; then, while the right hand twitched at the handle, the corresponding body clinging to the door edged itself round with a random attempt to close it. Then with a stagger the patient lurched forwards to our table with the gait you have seen just now, a tripping staggering gait, hastening and stopping, until he reached the chair. Yet even in the chair you see, as we did, that the man is still in incessant restless motion, like a marionette; now jerking an arm, now the trunk of the body, now shrugging a shoulder, and so on, just as a girl does in St. Vitus's dance. We are told that these movements never cease except in sleep. He is not grimacing, he puts his tongue out fairly steadily, and his speech is unaffected; notwithstanding, the first glimpse of the patient should suffice to stamp his malady on the mind of the observer.

X. Y. lives in a village in this county; and I am much indebted to Dr. Sydney Williams of Meldreth for the trouble he has kindly taken in obtaining careful and complete notes of the case and in arranging for the patient to attend at the hospital this morning.

X. Y., aged 46, was born in our county. He joined the army and served in India for seven years. Then he served in S. Africa three years. His father died at the age of 80; always a healthy man. His mother died at the age of 55; cause unknown. They had three sons, all living, and the two others are in good health; one, aged 51, is an officer's servant, the other, aged 48, an agricultural labourer. There is no history in the family of "fits, St. Vitus's, or insanity." In respect of personal history patient had always enjoyed excellent health, and escaped all infectious diseases—in his own words "he never knew his own strength." He is a life teetotaler, and never had any venereal disease. In India he took part in athletics, and ran the half mile for his regiment. He smokes about half an ounce of tobacco a week.

When on duty one night at the front with a pioneer battalion, as Zeppelins were about, orders were given to man the trenches; and in making for the trench he fell into it—some 6 or 7 ft.; he fell on his right knee, but did not regard the blow seriously till a month later when the involuntary movements began on the right side. Within two months the disorder had attacked the left side also, and soon all his body and limbs were involved in it. There is no apparent mental defect, but the patient says that his "memory has suffered much." He cannot retain his urine as well as he did. There is one point, almost the only one, in Huntington's chorea in which the movements

differ a little from ordinary chorea, namely, that in the earlier stages of the malady at any rate—as other observers of these cases have recorded—certain concerted movements may be performed with normal co-ordination; especially if the patient is tranquil and bent upon some task. Dr. Williams writes:—"I have noticed him in the road collecting dung for his garden, and when carrying the bucket and shovel the choreic movements are practically in abeyance." This occasional co-ordination, I repeat, agrees with records of other observers of this disease. One patient is said to have continued his occupation as a carpenter and to have "driven nails with fair precision." On the other hand, writing was almost impossible. The patient after much effort did succeed in making a sort of signature of his name, but it was scrambling and barely legible. The strength of his muscles seems unimpaired, at any rate as yet, and the tendon jerks were lively.

In short, this case corresponds in all respects with the recognized pattern of the malady, save as regards the absence of heredity. When heredity is apparent the malady has always been present in the preceding generation. There is a chorea described as senile chorea which comes on between the ages of 55 and 80, and is not hereditary. Other maladies affecting elderly persons, disorders of the nervous system, have been indiscriminately called chorea; such as athetosis, paralysis agitans, senile tremor, hysterical rhythmic spasms, and so forth. We can scarcely accept a limitation of the nomenclature by the element of heredity alone; but its absence may suggest a somewhat less gloomy prognosis. The incidence of Huntington's chorea falls rather more upon males, and usually about the age of 40.

Whether Huntington's chorea will prove to be identical with "senile" chorea I cannot say; we know too little of their respective natures and causes, but provisionally they should be regarded as one malady. It is said that both symptoms and morbid anatomy in Huntington's disease and in the "senile" are identical; though it is true that the occasional recovery from the "senile" kind, and perhaps the better preservation in it of the mental faculties, seem inconsistent with a parenchymatous degeneration attacking the neurons primarily, or very early. But these cases of alleged recovery are as yet imperfectly verified. Moreover we must not give the name of chorea to the common tremors and shuffles of elderly people with arterial and other senile degenerations. In these the motor instability is of another order. The descriptions of "senile" cases too often contain other forms of disorder, such as post-hemiplegic spasms, athetosis, shaking palsy, tics, festination, myoclonus, etc. The patient before us is no degenerate. It cannot be too clearly understood that in Huntington's chorea the muscular eccentricities are of the same order indeed as in Sydenham's chorea. By these alone the two kinds cannot be discriminated. One is tempted to say that all chorea proper results from interference, temporary or persistent, in the same (Rolandic?) area.

The last case of Huntington's chorea I saw was in Tunis. I happened to be at the window of my hotel, which looked on a narrow lane, when I saw a native, evidently afflicted with this malady, shuffling along and edging himself for support against the opposite wall. The movements as he walked became more and more violent, until, feeling himself shaken off his legs, he backed into a doorway, where leaning against the doorpost he scuffled down into a sitting position; there the poor fellow sat twitching, jerking and grimacing until in a few minutes the violent phase passed off; then, scrambling to his feet, he staggered off. I followed after him but failed to catch him. Accordingly I asked X. Y. if at times the agitation became such as to shake him off his legs, and he and his son—without whom he does not venture abroad—said at once that such was often the case with him.

The prognosis, even if the case be not inherited, is very grave; the muscles of mastication and speech become affected, and gradually, after many years it may be, the mind falls into degeneration, so that after many years, directly or indirectly, it proves mortal, the last phase being one of utter wreck. If in this case the fall were the cause of the attack, the outlook may be better than in a case of "spontaneous" origin; the symptoms may abate, and the tissue changes be arrested. No remedies known to us are of any value; scarcely even palliative. The

pathologists who have had the opportunity of examining the nervous centres after death tell us that distinct marks of feebly inflammatory decay are not inconspicuous in the brain; the spinal cord being normal. Especially in the Rolandic area signs of a slow pia-arachnitis are found, with degeneration of the cortical cells and some chronic cell proliferation. There are no affinities of this disease to rheumatic fever, but it would seem to be a result of some infection. In this case, as in others of the kind, the heart is unaffected. The causes of the malady are in complete obscurity; the discussions of its antecedents in the textbooks are trivial.

REFERENCES.

- ¹ *Medical and Surgical Reporter*, Philadelphia, 1872, xxvi, 317.
² In England by Suckling, Midland Medical Society, 1889.

THE NECESSITY FOR FULLER CONSIDERATION OF THE LOCAL PROCESSES OF DISEASE AND REPAIR IN THE TREATMENT OF PULMONARY TUBERCULOSIS.

BY

H. MORRISTON DAVIES, M.D., M.C.CANTAB., F.R.C.S.,
CAPTAIN R.A.M.C.(I),

CONSULTING SURGEON, UNIVERSITY COLLEGE HOSPITAL AND CITY
OF LONDON HOSPITAL FOR DISEASES OF THE CHEST.

THE recent discussion at the Medical Society of London, together with the leading article in the *BRITISH MEDICAL JOURNAL* of December 8th, 1917, have served to focus attention on the fact—which has been becoming increasingly plain for some time—that the sanatorium treatment of pulmonary tuberculosis is not fulfilling all the hopes of those who believed that in it a really satisfactory treatment had at last been attained, and that by it the therapeutic problem of consumption had been definitely solved. The method has had a long and thorough trial, but it must now be recognized that the results of it are not such as to justify us in continuing to regard it as a standard method, adequate to deal with the disease with reasonable success, and without radical improvement or addition. We cannot say that in the average case treated by sanatorium methods complete and permanent cure is the normal event. In any given case the result of the treatment is uncertain and extremely difficult to foretell; the immediate mortality remains considerable, the ultimate mortality large, while our power of completely arresting the disease and of restoring the patient to active life is still disappointingly capricious. While we are able in a large number of cases to produce the most gratifying improvement in the general condition and diminution of the symptoms, it must be admitted that the tendency to the persistence of some few signs or symptoms and to ultimate relapse after the resumption of working conditions are disappointingly common.

It is the object of this paper to inquire into the significance of the comparative failure of sanatorium treatment and to indicate some principles which that treatment ignores, but which must be taken account of before we can hope to establish a method in any sense definitive.

GENERAL PATHOLOGICAL CONSIDERATIONS.

In dealing with the problems presented by the treatment of pulmonary tuberculosis it is necessary to keep in mind constantly and vividly the general pathological features of the disease. Certain broad and quite elementary features of the processes involved in the disease will therefore be recalled. The application of these facts is so obvious, and seems when written so helpful, that no apology will be made for giving some account of them here. A familiarity with the pathology of consumption is usually taken for granted, and is therefore but little referred to in discussions upon treatment. A critical consideration of such discussions will, however, frequently reveal evidence that such pathological elements are, in fact, not always kept steadily and clearly in mind, and even that the appreciation of them is sometimes imperfect or confused.

The tubercle bacillus, when it infects man, shows itself to be an organism of slight general toxicity and slight local virulence. It does not in itself produce marked constitutional symptoms and the fever that it so commonly

causes is benign even when considerable in degree; similarly, the presence of the organism in the tissues has remarkably little direct destructive effect (as compared with the pyogenic organisms, for example), but rather stimulates them to produce a special tissue, the function of which is to surround and destroy the invading micro-organisms. This special tissue is, of course, granulation tissue and constitutes the well-known granuloma of tuberculosis. It is essential to grasp clearly the peculiarities of this tissue. It has no feature peculiar to tuberculosis, but is the normal response of the body to a mild continuous irritation. The function of the granuloma is to dispose of the irritant by destroying it, and its destiny, like that of all other granulation tissue, is, when its work has been accomplished, to be converted into fibrous tissue. If a few tubercle bacilli be introduced into a healthy tissue, the normal sequence of events is the formation of a granuloma round the bacilli, the destruction of the latter by the granuloma, and the direct development of the latter into a small healthy scar. Now we can define with fair precision the conditions necessary to ensure this sequence of events: (1) The general resistance of the patient to tuberculosis must be good; (2) the dose of the organism must be reasonably small; and (3) the part infected must be kept under certain local conditions known to be favourable to cure, of which by far the most important is a state of rest. When these three requirements are not satisfied the granuloma as a protective mechanism fails and becomes itself an agent of injury. Supposing such a failure to occur either from low general resistance, largeness of the dose (or what may virtually be the same thing, exceptional virulence of the organism), or unsatisfactory local conditions, the result is a progressive enlargement of the granuloma.*

This has two important consequences:

1. The granuloma enlarges at the expense of the surrounding healthy tissues, which it attacks and destroys, as it were to make room for itself, with a vigour approaching that of a malignant tumour. This is the mechanism whereby tuberculosis produces its destructive effects, which are then seen to be part of the actual defence against the disease. As is well known, the destructive effect of the enlarging granuloma is not equal against all tissues and is apt particularly to be restrained by any dense fibrous membrane and by the parietal pleura—a feature which is one of the few absolutely characteristic of tubercle.

2. The granuloma from its very nature draws its blood supply chiefly from capillaries, any other vessels it may receive being very imperfectly developed. The consequence is that when the granuloma reaches any considerable size the central parts become very feebly nourished and therefore undergo fatty degeneration with consequent caseation and liquefaction. As long as caseation has not occurred, direct conversion of the granuloma into fibrous tissue is still possible under favourable conditions; recovery with loss of tissue but without loss of substance then takes place. When caseation has occurred there is necessarily loss of substance as well as loss of functioning tissue. There is thus actual cavity formation, and recovery is no longer possible without diminution in volume of the affected part. When the irritation caused by the micro-organism is very mild the tendency to the direct conversion of the granuloma into fibrous tissue is very strong and is in action continuously, even though the disease is progressing. The lesion then much more closely resembles the syphilitic gumma than the traditional caseous tubercle. This continuous fibrous transformation is probably the mechanism that leads to fibrous phthisis and the fibrosing tuberculosis of lymphatic glands.

The fibrous tissue into which the successfully resistant granuloma ultimately is transformed tends, of course, to contract and to diminish the volume of the affected part, even although there may have been little or no caseation and loss of substance. Diminution in volume of the lung is thus seen to be an essential feature of recovery from phthisis; it is, as we have seen, rendered necessary in two ways—through actual loss of substance of the organ in consequence of caseation, and through the contraction which must accompany the conversion of granulation tissue into a mature and healthy scar. So essential a part

* I set aside for the moment the possibility that the failure of local control of the disease may be so complete that a generalized tuberculosis occurs.