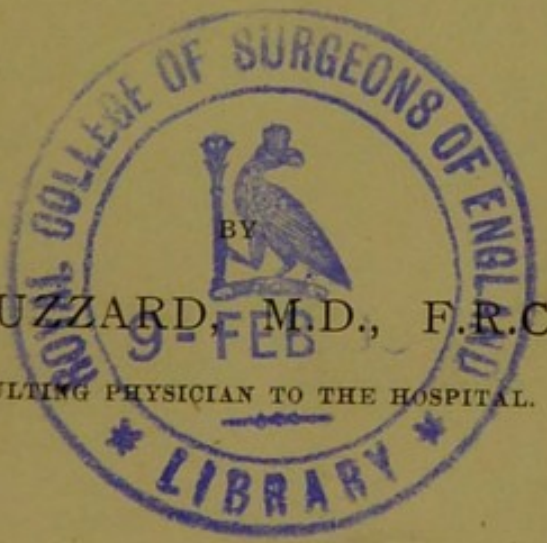
**A clinical lecture on remissions and relapses in insular sclerosis / by Thomas Buzzard.**

# A Clinical Lecture

ON

# REMISSIONS AND RELAPSES IN INSULAR SCLEROSIS

*Delivered in the National Hospital for the Paralysed and Epileptic, Queen-square, on Feb. 9th, 1904.*

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# A Clinical Lecture
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GENTLEMEN,—IT is known that in a very considerable proportion of cases the course of insular sclerosis is marked by more or less frequent remissions and recurrences of symptoms. The fact was recognised by Charcot and has been mentioned also by succeeding writers, but to my mind sufficient prominence has very seldom been given to this interesting and exceptional feature of the disease. It has appeared to me worth while, therefore, to devote a lecture to the subject and thereby perhaps to aid in drawing more particular attention to a peculiarity of the disease which may be found to bear with some importance upon its diagnosis and it is conceivable also upon the question of its etiology. This feature is illustrated by the case of a female patient in attendance.

The patient is a married woman, aged 36 years, who was admitted into the hospital under my care in September, 1900, complaining of loss of power in the left leg, weakness across the loins, a general feeling of weakness, numbness of the fingers of the left hand, and weakness of the bladder. Such were the symptoms which she herself described. Others were brought out upon examination, and we shall come to them presently. She said that her illness had lasted about 18 months, but on further questioning her it appeared that this short period by no means really represented the duration of her disorder. 12 years previously, in September, 1888, one year before her marriage, she had complained of numbness in the soles of the feet which "felt as if they were asleep." This lasted for a week and then passed off entirely. In September, 1889, about the time of her marriage, numbness in the feet re-

curred and this time it extended to the calves of the legs and lasted for about three weeks. Again it passed away completely. In August, 1890, when pregnant for the first time, she again suffered from numbness, which now extended from the knees to the nipples, and about the same time she had double vision and a "contraction of the left side of the face," described by her medical adviser as Bell's paralysis. These symptoms lasted about a month and then quite cleared off under treatment. In 1892, just as she was pregnant for the second time, she again had an exactly similar attack upon the left side of the face, which also lasted for a month. About the same time she likewise noticed some numbness in the fingers of both hands. This has never quite left her since, but has varied in degree from time to time. With the exception of this numbness, which was so slight as to cause her no practical inconvenience, and indeed scarcely to attract her attention, she remained completely well till December, 1895, a period of some three years. She was then confined for the third time and three or four months later had to seek medical assistance on account of numbness and loss of feeling in the hands and arms, so that "she could not sew, or peel potatoes." She also had some loss of power in the left knee and for two months was unable to walk. After being away in the country for a time she became practically well again in all respects. And so she remained until April, 1899, when, being again pregnant, she complained of heaviness in the legs and some weakness across the back, but no numbness on this occasion. The child, a healthy one, was born in September, but after her confinement she continued to become weaker and to complain of a "heavy pain" across the loins. At the same time she first began to notice some precipitancy of micturition. In 1900 her walking power diminished and she fell and was unable to draw up her left leg, the ankle and knee being stiff, and on admission here she could not walk without assistance. The bladder trouble, too, had been worse and there had been occasional incontinence of urine. There had been no further diplopia. The following note refers to her condition in November, 1901, when she was shown by me at a lecture in this theatre: "The patient is well nourished, of placid, easy-going disposition, cheerful, but not especially emotional. The bright but rather vacant expression is very suggestive of that which is so often noticed in cases of insular sclerosis, but mentally she is intelligent, with good power of memory

and attention. There is no articulatory difficulty. The ocular movements are good, but extreme lateral rotation is not well sustained to her right and when the eyes are turned to her left a few slow jerking movements are remarked, of a nystagmoid character. The jaw movements are equal and powerful on the two sides. There is no affection of sensibility in the face. There is no abnormality in the districts of the ninth, tenth, and twelfth nerves. In the upper extremities we find all movements powerfully performed and it may be said that there is no marked intention tremor, but when, by request, she touches the tip of her nose it is noticed that there is a slight wavy motion of the finger as it nears the face. In the lower extremities we find on the right side all movements present and power good. There is a little incoördination but no rigidity. On the left side the muscles are equally well developed but their power is much less and there is a great deal of spasticity. Dorsiflexion of the ankle is practically absent. Her gait is spastic, especially in the left leg, which is moved much like a hemiplegic limb. There are subjective numbness in the fingers, especially of the left hand, heaviness and dull pain across the loins, but no definite girdle sensation. Objectively there is no change to tactile stimulus. There is marked loss of sensibility to pain in the left leg as compared with the right, not quite constant, and extending from the foot (not including the sole) to above the knee without any definite limits. As regards reflex movements there is brisk jaw-jerk, the supinator jerk is in excess on the right side, the triceps jerk excessive on the right, absent on the left. The knee-jerks are both equally in excess and ankle clonus is present on each side. The plantar reflexes are both brisk and show extensor (Babinski) response. There is precipitate micturition with occasional incontinence of urine. Examination with the ophthalmoscope shows both discs rather pale, especially in the temporal half of each, and the left is more pale than the right ; the vessels are of good size. The field of vision, both to colours and light, is somewhat restricted concentrically in the left eye."

The patient for the last two years has been at home but attends to-day at my request. Her walking power since the above note was taken has much diminished and there is now some spasticity in the right lower extremity as well as in the left.

I have entered into what is perhaps a somewhat wearisome detail of this case because it is one which manifestly strays

from the commonly recognised type of insular sclerosis. Yet it is clear that this affords the only reasonable solution of the symptoms shown in the patient's history and present condition. In no other way can we explain the presence of such symptoms as transitory numbness, diplopia, short-lived but recurrent facial paralysis, loss of power with spasticity of gait, ankle clonus, the Babinski phenomenon, and pallor of the optic discs. This, then, is a case of insular sclerosis which well illustrates the importance of recoveries and relapses as a symptom of the disease. There is no doubt that not only have recoveries such as have occurred here been in many cases the cause of a patient's illness being lightly treated as one of hysteria but also, in my judgment, the common picture of hysteria has been somewhat largely based upon a misinterpretation of such symptoms.

Another example is that of a man, aged 35 years, tailor, married, who was first admitted under my care in April, 1895, with not strongly marked symptoms of insular sclerosis. It seemed that in 1881, when 21 years of age, he suffered from some hemianæsthesia from which he rapidly and completely recovered and remained well till 1894. He then complained of numbness, tingling, and weakness in both hands and feet. From this he recovered in a fortnight and returned to his work. About a year later he was again attacked with numbness in both feet and slightly in the hands, together with some weakness in both lower limbs, especially the right. There was also some slight bladder trouble. He had to give up work and was eventually admitted into this hospital. He had then distinct weakness in the right upper and slightly also in the right lower limb. There was loss of sensation, both to touch and pain, in front of each thigh. The knee-jerks were in excess. After a few weeks he had improved and was able to leave the hospital, walking alone. After his discharge he continued to improve and was apparently quite well in a month or two. He remained well and steadily at his work for nearly three years. In 1898 he began to be weak in the legs again but remained at work till June, 1900. He was then much worse and could not walk without help but after three weeks' stay in a convalescent home he improved greatly and returned home, but still continued to suffer from numbness and stiffness of the fingers which interfered with his work. He again became worse and was readmitted into this hospital in January, 1901. I will only refer to a few salient points of his

condition at that time. There was a little "intention" tremor in the arms, especially on the right side, and he was clumsy in both hands in picking up a pin. The right leg was weaker than the left, especially in dorsiflexion of the foot, and there was some rigidity on both sides, with incoördination of the right foot. His gait, too, was unsteady and of the spastic-ataxic type. The wrist and knee jerks were in excess, but there was no ankle clonus. The plantar reflexes were brisk, with extensor (Babinski) response. Patches of anæsthesia were found about his trunk and legs. This patient steadily improved in hospital at first, and the anæsthesia practically disappeared. After being discharged he again had a relapse and was unable to walk. There were marked increase in the spasticity of his legs and some return of anæsthesia. After keeping in bed for a short time he improved, but still showed an ataxic gait with some spasticity. He was discharged in May, 1901. At the present time he is under treatment in another hospital. On inquiry I have ascertained that he cannot stand without support or guide his hands. Work has been impracticable for four years past. (By the courtesy of hospital colleagues other cases illustrating the subject were shown.)

It is evident that the extreme irregularity which characterises the distribution of the patches of sclerosis in different examples of the disease under consideration makes it quite unreasonable to expect the same degree of uniformity in the symptoms that we are accustomed to find when sclerosis is of the "system" type, as, e.g., in tabes. It is by comparison of numerous examples, some of them verified by post-mortem examination, as well as by consideration of the effect which a sclerotic patch will be liable to produce when occupying any particular locality, that the ability to classify such cases as these under the name "insular sclerosis" is attained. The "symptom-complex"—to employ the rather unwieldy expression borrowed from the German—or "syndrome," to use the term employed by the French—the peculiar aggregation, that is, of the symptoms, points, I think, conclusively to this explanation of the cases.

In the course of private practice I have met with many examples—some of which have been already published[1]—

[1] Simulation of Hysteria by Organic Disease of the Nervous System. Churchill, 1891. Clinical Lecture on Insular Sclerosis and Hysteria. THE LANCET, Jan. 2nd, 1897, p. 1.

of the tendency to remissions and relapses in insular sclerosis. Let me refer, though very briefly, to a few more of these, because in a subject of this kind a description of incidents occurring in concrete examples is more striking than any abstract statements.

CASE 1.—A woman, aged 33 years, whom I saw about a month ago, had an attack of influenza in 1895. Shortly afterwards she squinted and lost the use of her right hand and arm—i.e., although able to move it she could not use it for lifting things or dressing. She also lost feeling in it. At that time she dragged one of her feet, although she was not conscious of loss of power in it. The trouble in her limbs described lasted for about a month and then improved, although she did not quite regain the power. She had been very much improved for about a month when she relapsed and this time both right arm and leg were affected. For three weeks she was very helpless and stayed in bed for the most part. She then began to improve and perfectly recovered in the course of some months. From that time (between seven and eight years ago) she remained perfectly well until June, 1902, when, as she describes it, the right arm began to be weak and the weakness spread to the right hand. Just as happened before, she again became practically helpless in this limb and the leg was also affected to a certain extent. She went to the seaside and after a month had quite recovered. She remained well for a month and then felt her leg losing power again, and although keeping about she had great difficulty in walking. The right arm, too, was slightly weaker but she was able to use it. On the first attack eight years ago there was some difficulty in restraining the action of the bladder and this difficulty recurred in the last attack. There is now some weakness of the right limbs with attempted ankle clonus. On the occasion of my examination I failed to elicit any plantar reflex.

CASE 2.—A woman, now (1904) aged 48 years, lost the sight of her right eye in less than a week when 23 years old. At its worst she could not perceive light. It gradually recovered completely. A few years later she had transitory tingling in the right arm and weakness in one ankle which passed off. About 1894 there were loss of power to hold a pencil and dragging of the right leg, from which she also recovered. I saw her in 1898 when the right leg was very

useless and she could only walk with help on each side. In reply to my recent inquiry her medical attendant reports the legs spastically paralysed, especially the right, presence of the Babinski phenomenon, and very feeble grasp of the hands.

CASE 3.—A woman, aged 22 years, was in excellent health till February, 1899, when vision with the left eye became indistinct and she had headache and giddiness. The left hand and arm were numbed and she could not walk properly, the right leg especially being weak. In April, 1899, an ophthalmic authority found vision on the right side $\frac{6}{9}$, on the left $\frac{6}{60}$, both eyes emmetropic, and the fundi normal. The diagnosis given was "functional derangement of nerves at the back of the eye." In April, 1900, I found right vision = $\frac{6}{6}$, left vision = $\frac{6}{9}$. The fundi were normal. There was nystagmus. She complained of "pins and needles" and numbness in both hands and below the left knee; she could walk a mile but would then be tired. There had been inability to hold a pen but she could now write. The plantar reflexes showed extension (Babinski) movement. I thought that she had suffered from retro-ocular neuritis in the course of insular sclerosis. Under observation in London she greatly recovered her health and her medical attendant wrote me a year or more later, "I am glad to tell you that she is doing well, hunting, and driving about in her old dashing style, and they speak of her as being 'quite well now.'" A few days ago I heard that she had had no return of the numbness or loss of power in her limbs, and that she was able to take riding, driving, and walking exercise, though in riding she had not regained the full grip of the saddle. It is, I suppose, possible that there may be no new developments in this case, but in view of what so commonly happens one cannot be at all free from apprehension that a relapse may some day occur.[2]

[2] Since this lecture was sent to press I have had the opportunity of once more seeing this patient. She looks the picture of health, and is studying in a school of art. On examination I note the following. Her upper extremities are absolutely right; there is no numbness or "pins and needles" in them, and she is able to draw with a perfectly steady hand. If she walks much the "feeling seems to go in her left knee and leg." Two years ago she could dance well but of late this has been quite impossible, owing to extreme giddiness. Holding the head down causes giddiness. During last winter she hunted, but owing to her want of grip, especially with the left leg, in jumping she had to rely very much upon her balance instead of depending upon the

CASE 4.—A woman, in 1894, suffered from weakness and tremor in her legs, for which she had to keep her bed for some days. From this she recovered perfectly and remained well till 1897, when she began to lose strength in her legs and fell on several occasions. Four months later I found some spastic paraplegia, weakness of the sphincters, excessive knee-jerks, ankle clonus, and pallor of the left optic disc. The vision = $\frac{6}{12}$; there was some intention tremor of the hands.

CASE 5.—A woman, aged 31 years, whom I saw in 1901, suffered from an illness at 13 years of age, in which she had double vision and squinting, and staggered in walking. After two or three months she quite recovered, Three years later the sight of the left eye became bad and an ophthalmic authority who examined her is said to have expected that she would become blind; but great improvement occurred and the vision with that eye I found to be $\frac{6}{9}$, though the disc was distinctly atrophic. Diplopia had since occurred for several weeks at a time on three different occasions. A few years ago, however, she was so well that she walked ten hours a day in Switzerland. After this there was for a time some loss of power in the right leg. More recently the left leg became weak and still remained so when I saw her. She presented, besides the eye symptoms mentioned, a characteristic staggering gait, excessive knee-jerks, extension (Babinski) movement in the plantar reflex on both sides, and slight nystagmiform jerking. At times she had had numbness in her hands with inability to use them properly but that was not now the case. Some time after my examination I learned that she had again recovered her health and had married. I am unable to report her present condition.

CASE 6.—A woman, aged 36 years, was sent to me in April, 1899, suffering from loss of power in both legs, back, hands and arms, a staggering gait, and stiffness from the hip to the knee. The knee-jerks were excessive, ankle clonus was present on each side, with marked extension (Babinski) response in the plantar

crutch. The plantar reflexes are of the extensor (Babinski) type. During the last year or so there has been a return of the visual difficulty which, according to her account, had entirely subsided and I now find V. R. = $\frac{6}{9}$, L. = $\frac{6}{24}$ with paracentral scotoma. She has no diplopia. Both discs are pale in the outer part, especially the left.

reflex. There was nystagmus and also atrophy of the left optic disc. The history was that she had been particularly healthy, and exceptionally strong muscularly, until 1890, when she suffered from two attacks of influenza. After being in bed for ten days in the second attack there was some loss of power and sensation in her legs. After this, although she recovered her general health completely, she thought that she never seemed to regain the "spring" in her walk but at times she was so much improved as to be unconscious of anything being wrong. After this there appear to have been several relapses and remissions. At one time there was involuntary escape of urine and fæces but this passed off in six weeks. For one and a half years she had complained of diminished sight of the left eye and latterly that of the right had not been strong. During the nine years of illness her case had been always diagnosed as one of hysteria except by the last medical man consulted who recognised its nature and sent her to me. In 1901 I received from him the following in reply to my inquiry as to the sequel of the case. "The mother was so annoyed at me for saying it was not hysteria and at you for supporting me that she removed the case out of my hands. I have made inquiries among some of the relatives but can get no information except that the symptoms had slowly increased and she died of exhaustion about 15 months ago."

Here I am tempted to quote from some notes of a case which I took in May, 1868, at a time when this disease had been, I think, scarcely differentiated and when certainly I knew nothing about it.

CASE 7.—The patient, a man, aged 41 years, had never had syphilis or any severe illness or trauma. 23 years ago he found himself one day lame in the left leg with twitchings in it and a feeling as though it were asleep. In a fortnight he was quite well and remained so for three or four years. Then he had a second attack like the first which lasted also about a fortnight, when he returned to business. A few years later he had another attack of similar character and has never since quite regained the power of the left leg. Two years previously he felt some numbness of the left hand. He has never had pain in the leg but of late a little walking, even for half a mile, would exhaust him utterly. A few days previously he felt for the first time powerlessness in the right leg with numbness. There was also

diminished grasp of the right hand and for several days he had headache and lost power rapidly in his limbs and back. For the last few days he had been getting better again. In the morning his left leg was stretched out and he could not bend it. The grasp of the right hand was fair ; that of the left was defective. The extensors of the left toes failed to lift the foot voluntarily but tickling the sole caused them to act "fiercely"—reflex excitability was very much exalted. (The Babinski phenomenon was evidently present.) Sight had been hazy during the last few months. The expelling power of the bladder was not great and he could not hold his urine to a great extent. I do not know the sequel of this case which was evidently, however, one of the class which we are considering.

Case 8. To complete the picture let me now quote notes of a case which was under my care in this hospital many years ago the diagnosis of which was confirmed by necropsy. In May, 1885, a female, aged 27 years, previously in perfect health, fell upon her hands whilst running and rose with great difficulty on account of extreme giddiness which had doubtless occasioned her fall. In a few minutes she recovered and for a week had nothing to complain of, then her sight became indistinct, she had diplopia, and pain at the top of the head. She attended here as an out-patient and gradually lost her symptoms. In February, 1886, she was suddenly unable to walk and lost power also in her arms. Two months later she could not stand. There were difficulty in micturition and occasional leakage of the bladder. Admitted as an in-patient in July, 1886, she lay in bed with but little power of moving her limbs. There was some anæsthesia of the lower extremities. The knee-jerks and plantar reflexes were present ; there was no ankle clonus. Later she suffered from spastic rigidity of the legs, marked interosseal muscular atrophy of the hands, absence of knee-jerks, acute bed-sores, bladder trouble, and she died in November from gradual heart failure after constant vomiting. From first to last her illness occupied just 18 months. Post-mortem examination showed on the posterior surface of the cord just above the lumbar swelling beneath the pia mater a slightly raised, rather dark-looking patch. On each side of this was a black-looking vein filled with clot, contrasting with the almost empty pink-looking veins elsewhere. Section showed soft greyish-looking tissue, replacing in part the white and grey substance of the cord. Sections

through the lumbar cord showed in some parts of the anterior and lateral columns grey semi-gelatinous patches. In the mid-dorsal region the whole transverse area of the cord flowed out on section, having an opaque grey colour. In the mid-cervical region sections showed grey gelatinous patches occupying various portions of the axis.

In the present state of our ignorance regarding the etiology of insular sclerosis there is no advantage in disguising the fact that we are unable to point to any mode of treatment which appears to influence the progress of this disease. It becomes then a question of considerable importance whether the remarkable tendency to the occurrence of remissions and relapses in its course is calculated to throw any light on the mode of its causation. As it seems to me the most likely view (and possibly that which finds most favour) is that in insular sclerosis the essential cause is the presence in the blood-vessels of an infective agent which sets up an inflammatory process in the interstitial tissue. This may occur at any part of the cerebro-spinal axis. On the other haud, it has been suggested that the pathological process is essentially dependent upon some congenital abnormality. I am disposed to think that such an explanation must be set aside. The grossness of the anatomical findings alone would seem to be inconsistent with such an hypothesis. Besides this, insular sclerosis cannot be said to be a family disease, which might be expected were the disease of congenital origin. It is, in my experience, exceedingly rare to meet with more than one example in a family. Diseases of the nervous system of endogenous origin are usually characterised as well by the fineness of the changes observed in the structures involved as by their slow and comparatively steady progress, uninterrupted by any important tendency to marked remissions and relapses. On the other hand, if we turn to disease of exogenous origin we shall find that in one, undoubtedly and pre-eminently of infective origin—syphilis—we are apt to meet with this peculiar feature very distinctly exhibited. Thanks to the fact that specific treatment in syphilis has a powerful and very definite influence upon most cases of the disease, it is quite likely that the tendency to recovery, followed sooner or later by relapse, in that disease is a good deal obscured. But, even as it is, that tendency is constantly showing itself after a time when the beneficial effects of the

remedies may be assumed to have passed off. In syphilis we have to deal with comparatively gross changes—inflammation, gummata, endarteritis, and the like. For the most part the changes in insular sclerosis may be said to be of gross character. It is true that some observers have pointed to the fact that occasionally the anatomical appearances in insular sclerosis cast some doubt upon the more usually received opinion that the disease is of inflammatory origin. Redlich, for example, amongst others, has found in insular sclerosis nerve fibres profoundly affected along with relatively little-changed neuroglia, and says that vascular changes happen only in a certain number of cases. But, as I have pointed out on another occasion, this is by no means conclusive. Parenchymatous degeneration of the posterior columns in a case of tabes may be accompanied by gross intracranial endarteritis. Presumably, therefore, a toxin may give rise coincidently to each form, and in some cases of insular sclerosis—or in some localities in a particular case—its influence may fall upon the nerve fibres rather than on the vessels and interstitial tissue.

The remarkable variations in the degree of acuteness in insular sclerosis, again, appear to be much more consistent with an infective than an endogenous origin. I have had cases under observation in this hospital in one of which 15 and in another 19 months only had elapsed from the commencement of symptoms and death. One that I met with in private (which was not, however, verified by necropsy) lived only 12 weeks. Ribbert examined anatomically one that survived only for three months. More often, as is well known, a far longer period characterises these cases, which may drag on for ten or 20 years or more.

We are well accustomed to the differing degree of acuteness in all forms of infective disorder, due probably either to intensity of the infecting agent or diminished power of resistance. In Friedreich's ataxia, on the other hand, to instance what is probably an example of congenital defect, the course of the disease is practically uninterrupted and chronic in character.

I fail also to see adequate grounds for the suggestion which has been more recently offered that insular sclerosis is the result of a toxic influence upon an existing defect of development. The presupposition of such a congenital defect appears to me as unnecessary as it would be in the case of infantile paralysis or of the neuritis which so often is a sequel of diphtheria. The frequency with

which insular sclerosis attacks persons of exceptionally fine physique, the extreme rarity of its occurrence in more than one member of a family, as well as the marked remissions—often amounting to temporary recovery—which may take place in its course, appear to be inconsistent with such an explanation and to be much more in accord with the view that the disease is purely of toxic origin.

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