A CLINICAL ANALYSIS OF AN EXTRAPYRAMIDAL SYNDROME ; PARALYSIS AGITANS AND POSTENCEPHALITIC PARKINSONISM.

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INTRODUCTION.

THE extrapyramidal system is a complex system to lesions of which authors have ascribed different clinical conditions ; some of these ascriptions have stood the test of time, others have fallen into disrepute. The commonest affection of the system is paralysis agitans which has for its pathological basis lesions confined mainly to the corpus striatum and its efferent connexions.

In the Croonian lectures of 1925 delivered by S. A. K. Wilson disorders of motility and of muscle tone were considered at great length and with clear understanding. In these lectures paralysis agitans was used as part of his text. As a follow-up plan to that of the author of the Croonian lectures and to substantiate further the dicta laid down by him, in personal touch with him though working independently, I have collected and examined a series of cases of the syndrome from his services at the National Hospital, Queen Square, and at King's College Hospital.

GENERAL SURVEY OF CASES.

Fifty cases were selected, of which 39 were cases of the Parkinsonian syndrome of postencephalitic origin, while 11 belonged to the idiopathic type of the disease. Thirty-four males and 16 females made up the 50. Both sides of the body were affected equally in three cases ; the right more than the left in 20 cases, the left more than right in 18 cases, and the right arm and left leg in one case. The right side was alone affected in five cases, and the left in three.

The ages of the postencephalitics varied from nine years to the fifth decade, the majority (26 in all) occurring between the ages of 20 and 40. The fifth and sixth decade was the average age-period of the idiopathic cases.

The 39 patients with postencephalitic Parkinsonism gave an account of an *earlier illness*, diagnosed at the time as encephalitis lethargica in 29 instances, influenza in five, rheumatic fever in two, and appendicitis in one. One patient had had no previous illness.

The *symptoms at the time of the original illness* consisted of somnolence in 26 cases, diplopia in 19, delirium in 11, restlessness in six, headache in five, and giddiness in two.

The *interval* between the original illness and the onset of the Parkinsonian symptoms amounted to four years and four months as a maximum ; in 18 cases no interval was remarked. The average worked out at 7.2 months.

The *duration* of the Parkinsonism in the postencephalitics was seven years in the longest instance and three months in the shortest, making an average of two years and two months.

The idiopathic group of eleven cases gave for the longest duration of symptoms six years, for the shortest nine months, and an average 2 years and 11 months.

The *symptoms in order of frequency* characterizing the Parkinsonism of the 50 cases of this series consisted of stiffness in 45 (of which nine belong to the idiopathic group); shakiness in 37 (including ten of the idiopathic group); dribbling in 25 (of which two belonged to the idiopathic type); "weakness" in 24 (of which eight were idiopathic cases); and diplopia in 20 instances (of which four belonged to the idiopathic group). Postencephalitic tic occurred in four cases. Character change was remarked in two cases and delusional formation in one.

GENERAL ATTITUDE.

(1) *Standing*.—In eight cases, evenly distributed between the two groups, a normal attitude was assumed.

The other cases demonstrated the usual attitude of flexion, which varied in degree. The head was flexed on the shoulders, the shoulders were stooped, the affected arm bent slightly at the elbow, the arm adducted and the hands in a flexed position. The trunk was bent slightly forward and the legs bent at the knees.

(2) *Sitting*.—The attitude was normal in 16 cases, of which four were idiopathic.

In the other cases the patients did not use the back of the chair, the feet were tucked well underneath, the body was inclined forward and the gaze directed to the floor. In the severe cases the patients, as they sat down, would slightly flex their knees and then fall into the chair; at the same time the feet were moved out. While sitting they would appear very uncomfortable and stiff.

(3) *Expression*.—The expression was unaltered in five cases, of which four were the idiopathic type. The other patients exhibited a slight or a marked change in their facial expression, as indicated by the mask-like fixed facies, the partly open mouth, the greasy skin and the bright eyes. Numerous statements made by the patients commented on this change.

OCULAR CONDITION.

(1) *Accommodation* was good in twelve cases, of which four belonged to idiopathic paralysis agitans. It was fair in eleven cases, of which five were idiopathic; poor in ten cases (two idiopathic), and absent in 17 cases (all postencephalitic).

(2) *Light*.—The reaction of the pupils to light was always as good as, and in most cases better than, the accommodation reaction. It was good in 28

cases, of which ten were idiopathic. It was fair in 17 cases (one idiopathic); poor in four cases and absent in one.

(3) The pupils were unequal in three cases.

(4) *Convergence* was good in 13 cases, which include six cases of the idiopathic variety. In the others convergence was defective in variable degrees; some showed no power of convergence, others converged with one eye and not the other. Good convergence with a rapid swinging out of the eyes immediately after was also seen, and in some instances it was only partially restricted. When one side of the body was affected more than the other as regards the Parkinsonian syndrome it was noted that convergence was defective in the eye of the unaffected side in 14 cases, and in the eye of the affected side in four cases.

(5) Defect in *outward* deviation of the eyes occurred in 17 instances.

(6) *Blepharospasm* was present in 45 cases, nine of which were examples of idiopathic paralysis agitans.

(7) *Weakness of the orbicularis oculi* was found in 33 cases, seven of which were idiopathic.

(8) *Punctuate movements and "blinking."*—Punctuate movements consist of several jerky movements of the eyes on looking to the right or left with the head held steadily in front. By 'blinking' is meant a momentary closure of the lids as the patient looks from one side to the other; a synkinesis or associated movement first described by Wilson in his Croonian lectures.

Punctuate movements were present in 28 cases (three idiopathic) and absent in the remaining 22 cases. In ten of the cases which showed no punctuate movements the rest of the examination of the eyes was normal, but in the remaining twelve there was a defect in respect of accommodation, convergence and light reaction in varying degree. In no instance was the accommodation reaction entirely absent.

'Blinking,' in the sense mentioned above, occurred in 29 cases (three idiopathic) and was absent in the remaining 21. The 'blinking' corrected the punctuate movements in five cases. In the other cases it had no effect.

(9) *Micropsia* was found in two cases, both belonging to the postencephalitic group.

MOUTH AND TONGUE.

At rest, the lips were approximated in 31 cases, of which 11 cases belong to the idiopathic group; in the remaining 19 the mouth was always partially open.

Volitional movements of the mouth were normal in nine cases (four idiopathic). They were reduced in 41 cases; equally on both sides in eleven cases, right side more than left in 16 cases corresponding to the side of the body which was more severely affected, left side more than the right in 14 cases corresponding similarly to the side more affected.

On emotional innervation no variation from the normal was noted in ten cases, of which four were idiopathic. Of the remaining 40 cases the right side was affected more than the left in nine instances and *vice versa* in seven. The side of weakness corresponded to the side of the body affected by the Parkinsonian condition.

The range of movement of the tongue was reduced in 30 cases, of which only two belonged to the idiopathic class. Tremor of the tongue was present in 34 cases, seven of which were idiopathic. Tremor of the lips was present in eight cases, and in two a fine tremor of the cheek muscles.

SPEECH.

Articulation and phonation were normal in 21 cases, three being idiopathic. The other 29 patients exhibited little or no modulation to their voice, which was monotone in character. A tendency to stammer was noted in some, with slurring over the words and variation in rate, at times slow and then again very fast. Often the words would come slowly at first and then the speed would increase, with the result that the words would be run together. The monotone character was very noticeable. "It seems as if I was always talking on the same scale." "My voice is not so musical." A tendency to tire was also pronounced, and often a subjective observation. Sometimes the patient could not make himself understood. Palilalia was noted in one instance in the repetition of "No, no, no," or "Yes, yes, yes," etc.

Micrographia was present in 14 cases, of which two were idiopathic. The script was smaller and closer together, and very shaky. In two cases writing could not be performed because of the affection.

MOVEMENTS OF COOPERATION.

Arm-Swinging in Walking.—In two cases only was the swinging of the arms in walking normal. In everyone of the other 48 cases it was defective. This defect consisted in absence of swinging of both arms in 24; absence of the swing of the right arm in seven; absence of the swing of the left arm in eight. It was partially diminished in both in three; partial swing of the right was noted in four, and partial swing of the left in two.

With volitional effort 20 of the patients was able to swing their arms, but this always appeared to be laboured and to have no useful influence on the walking. It did not seem to have any co-operative action. The side more affected showed less excursion of the arm on volition than the other side.

Remarks made by the patients were as follows:—"They won't swing." "I seem to keep my left arm in one position. It seems awkward to swing it." One patient when asked to swing his arms remarked "I can't," and made a very poor attempt. Others said, "It is very difficult to swing the arms." "I forget to swing them."

Sitting to Standing.—In rising from the sitting position the feet are placed well under the centre of gravity and the body bends forwards as the subject

swings up to the standing position. In 32 cases this movement was normal; the feet moved in and the body bent forward. In ten cases there was no movement of the feet, but since they were already placed well under the patients, owing to the attitude of flexion, it was really unnecessary to move them.

Standing to Sitting.—As a rule when one sits down the feet move out. In 23 cases this movement was performed as in the normal. In 27 cases there was no movement of the feet after assuming the sitting position, the feet remaining well under the patient. Of the 23 cases where the feet moved out, in four the feet sprang out as the patient fell into the sitting position. In five cases one foot moved out and not the other, the foot which moved being always that of the side less affected.

ASSOCIATED MOVEMENTS.

The movements of association examined were abduction of the little finger, abduction of the thumb, flexion and extension of the fingers, and pronation and supination of the hand. As a rule these movements of association, when present, were obtained from the affected to the sound side.

In 26 cases in which all the above associated movements were present they occurred from the right to the left side in 13 instances, in all of which the right side was more affected than the left; in 14 cases the associated movements were from left to right, all of which were left-sided cases, with the exception of one which was a right-sided case and showed associated movements from either side.

In nine cases these associated movements were entirely absent though in other respects these nine were identical with the other cases examined in respect of motility and tone.

The reason for the absence could not be ascertained.

In 15 cases the associated movements were present in varying degree. In ten abduction of the little finger in association with abduction of the corresponding finger of the opposite side was present; in seven abduction of the thumb was the associated movement; in five, it consisted of flexion and extension of the fingers; and in nine of supination and pronation of the hand.

The associated movement was from the affected to the sound side in all these cases, with the exception of two where it was from the sound side to the affected. In three cases it occurred from either side, though in each the right side was more affected than the left.

SIMPLE AND COMBINED MOVEMENTS.

- (1) Shut the eyes with or without resistance; eyeballs roll up. This combination was present in the 50 cases.
- (2) Turn eyes to the side; head turns also. Reduced in six cases, but never absent.
- (3) Open mouth against resistance; head extends. Absent in two cases, reduced in one.

- (4) Turn eyes up ; forehead wrinkles, head extends. Absence of wrinkling of the forehead in nine cases ; reduced in seven. Head extension present in all cases.
- (5) In supine position, flex head on chest ; recti abdominis contract. Present in all cases.
- (6) Abduct extended arm at shoulder ; opposite erector spinæ contracts. Present in all cases.
- (7) Flex fingers in grasping ; extensors of wrist contract. Present in all cases but reduced in four.
- (8) Open closed hand ; extensors of wrist relax ; flexors of wrist contract. Present in all cases.

GENERAL MOTILITY.

The actual *muscle power* in paralysis agitans expressed subjectively and as found by objective examination is decreased in a special manner. The bigger movements, such as flexion and extension at the elbow and movements at the shoulder are proportionately much more forceful than smaller movements carried out by the hand. In those cases where the tone was normal or where tremor was lacking the muscle power tended to be good. In eleven cases no objective decrease in strength could be ascertained by testing the grip, or in abduction and adduction of the fingers, or in movements at the elbow and shoulder, though it must be remarked that some of these cases showed an increase of tone and some tremor. In one of these cases the patient complained of a subjective weakness.

In 39 cases weakness of the limbs was present especially in the movements of the small muscles of the hand. In these cases it was noticed that a position could be maintained against strong resistance for some time. The static strength was good but when a movement was attempted against resistance weakness was observed. The weakness was often put down to the tremor of the patient. "I feel stronger in the right hand as this one (left) is so shaky." "It is not weakness. It is really the shaking of it." Often the weakness was expressed as fatigability. "My left hand tires more quickly than my right." Sometimes the patient could not define the weakness. "I don't know whether it is actually weak. I don't feel to have the strength." "My hand seems peculiar. It does not act so well as it ought to." In other instances the weakness was expressed as due to the stiffness. "My hands feel stiff, not particularly weak." "My hands don't feel weak. They are stiff." Sometimes the weakness was the result of the slowness of movement. "I can't say they are weak. It is the slow movement."

The *range of movement* was limited on the affected side in 44 cases. This limitation was much more pronounced in smaller movements than in bigger ones. Abduction of the fingers gave a greater percentage of decrease than any other movement. "I cannot do fine movements with my left hand. It is from the wrist down and in my legs from my ankles down." Next in order of

frequency of limitation was approximation of thumb to little finger. The big movements such as flexion and extension at the elbow were very infrequently affected, this defect occurring in 18 cases. "I am able to do bigger movements better than smaller ones." "I can box or do anything active." "I can take a longer step better than a short one." "I can run better than I can walk."

Freedom of movement was examined by questioning the patients as to their ability to shave, feed themselves, cut their food and perform the ordinary routine things of life. It was noticed that they made very infrequent movements and that their movements of defence were slow in beginning and retarded in speed.

In 41 cases freedom of movement was reduced, the patient often favouring the unaffected side to do his routine, such as using the left hand instead of the right to shave himself. Poverty, slowness and lack of freedom were attributed by some to shakiness. "I am frightened to get hold of anything because they shake so." In other instances the alteration was explained as the result of the increase of tone. "My hand seems to become fixed when I use it." "I am able to carry out very quick movements better than slow ones." "I can run up a hill more easily than I can walk up." "If I get up with a jerk I can get up."

Fluctuation of strength, range, freedom, tremor and tone was volunteered as a personal observation in 24 instances. "Sometimes I am not so bad as others. I don't shake." "At times I get into a slow way. Sometimes I am quite agile." Several patients had the feeling that they were better in the morning; at other times it was just a daily variation. During warm weather a few patients felt better. Four patients expressed a feeling of ease of movement towards evening.

Restlessness.—In 18 cases a feeling of restlessness was pronounced even though the whole stance of the patients suggested immobility. When washing and drying themselves they had to keep on the move. They became restless after reading a few moments. They simply had to keep "on the go." A patient's wife expressed this very well: "He drives me mad at home—up and down all the time." Another patient said, "I can't sit still. My wife is always grumbling about it."

Tone.—Increase of tone was very common, occurring in 43 cases; ten showed a slight increase of tone and 33 a great increase. This hypertonia was tested by passive extension and flexion of the arm at the elbow. In the movements of the smaller joints the alteration of tone was less easily recognised.

In seven cases there was no perceptible alteration, though three of these patients showed a decrease in muscular power; three, a limitation in freedom of movement; three, a defect in range. Six exhibited an obvious tremor.

TREMOR.

In 45 cases tremor was present, consisting either of an action-tremor or a tremor at rest. It varied considerably in range, speed, and localization.

Action-tremor, or tremor on movement, was observed in 15 cases. When any movement with the affected limb was made the tremor would be increased, as in the act of shaving or of cutting food.

Tremor at rest was present in three cases. Both types of tremor was present in 27 cases; action-tremor was more pronounced in 18 and rest tremor more active in nine. Rate and range varied within wide limits from a very slow, wide excursion tremor to a fine, quick tremor. In 14 cases the tremor could be said to be wide in range and slow in rate; in the other 21 it was fine and rapid. Often the tremor was fine at rest and would become very coarse and irregular with volitional movement. Frequently the tremor at rest was observed to decrease at the commencement of movement and to become accentuated as the movement came to termination. An effort of any description, such as forceful closing of the opposite hand, often greatly increased the tremor.

The tremor could be controlled in many instances by slightly different ways, *e.g.*, by pressing the back of the hand on the chest, when only the thumb which was free showed the tremor, or by holding the palmar surface of the hand flat on the chest, or by gripping the hand with the other. On supporting the arm the tremor was greatly reduced in a few cases. In one case any change of posture of the limb would stop the tremor momentarily, and then it would begin again with the limb in the new position. Gentle support of the fingers would often check it. Under emotional stimulation it increased. Concentration on the tremor often caused a cessation of it for a brief moment.

The tremor was chiefly confined to the arms, for it was observed in 45 cases in some part of the arm, *viz.*, 21 cases in arm, hand and fingers, and 24 in hand and fingers. The legs were less often affected, this occurring in 15 instances. In two cases the cheeks showed a fine tremor.

The idiopathic cases of paralysis agitans all exhibited increase of tone, weakness, limitation of range and poverty of movement, and tremor. The tremor was variable in character, action-tremor being more marked in five cases than tremor at rest, and in six tremor was reduced by any voluntary movement.

Of the five cases in which tremor was lacking both at rest and on movement, strength was reduced in the grip in three cases, range of movement reduced in three, freedom of movement altered in three, and hypertonia found in four.

COMPARISON OF IDIOPATHIC AND POSTENCEPHALITIC CASES.

Although the neurologist meets with cases of Parkinsonism in young people from whom no history of encephalitis, or even of influenza, can be obtained, and legitimately assigns these to an unrecognised encephalitic condition, I have taken as idiopathic paralysis agitans the eleven of this series largely because of the ages of the patients and the extremely slow progression of the symptoms. In none, needless to say, was a history of encephalitis forthcoming.

A comparison of the symptomatology of the two classes is not without interest, especially in view of the contention of some writers that they are not in reality distinguishable. Taking first the symptoms complained of, in order of frequency, I have worked out the percentages for the two classes and embodied them in the accompanying table.

TABLE I.

					Postencephalitic group.	Idiopathic group.
Stiffness	...	...	...	...	92	81
Shakiness	...	...	...	...	69	91
Dribbling	...	...	...	...	59	18
Weakness	...	...	...	...	41	72
Diplopia	...	...	...	...	41	36

Making allowance for the fact that the numbers of the idiopathic group are but one-third of the other, I think nevertheless that the distinctions are definite enough to be of value. Thus shakiness or tremor occurs with considerably greater frequency among the idiopathics than among the postencephalitics, while stiffness occurs less often. These figures bear out the contention of most clinicians that the idiopathic class usually shows more tremor and the postencephalitic class more rigidity. Again, sialorrhœa is much more noticeable in the latter group than in the former. 'Weakness,' on the contrary, characterises the idiopathic group as a subjective complaint to a greater extent than the postencephalitic. In respect of double vision there is no material difference.

Proceeding next to the results of objective examination, I have collected them statistically in the adjoining Table (figures expressed in percentages).

TABLE II.

					Postencephalitic group.	Idiopathic group.
Accommodation, poor or lost	...				64	18
Convergence, poor or lost	...	...			81	54
Pupil reaction to light, poor or lost					13	0
Blepharospasm	...	...	...		92	82
Orbicularis oculi, weakness...	...				67	63
'Blinking' sign	...	...	...		67	27
Tongue movements, poor or weak					72	18
Tongue tremor	...	...	...		69	63

From this comparison it will be noted that blepharospasm is the commonest of all the objective changes in the class of ocular, facial, and tongue movements, and that its percentage is identical with that of the rigidity given in Table I. Next in frequency comes defect or loss of convergence, which is considerably more common among the postencephalitics than among the idiopathics, and the same is true of accommodation. Weakness in power and poorness of range of tongue movements are similarly frequent in the case of the postencephalitic variety, being no less than four times as common as in the case of the idiopathic group. Tongue tremor, on the other hand, is approximately equal in frequency in the two.

In fact, the general conclusion to be drawn from the figures given in Table II is that in respect of every one of the objective signs there enumerated its occurrence is more common in the cases of Parkinsonism following epidemic encephalitis than in the idiopathic class. Only as regards tongue tremor, weakness of the orbicularis oculi, and blepharospasm do the two approach each other.

As for the general symptomatological condition in regard to rigidity, hypertonus, muscle power, and also in respect of the special motor tests which have been employed, no material difference between the two groups has been established.

GENERAL CONCLUSIONS.

My clinical analysis of 50 cases of the Parkinsonian syndrome enables me to draw several conclusions which corroborate the observations made by S. A. K. Wilson in his Croonian lectures and substantiate the important deductions which he has there drawn.

In testing the synergic action of muscle groups in Parkinsonism, I have failed to find any departure from normal, physiological, law. In all cases, without exception, there is no interference with the appropriate interaction of protagonist, antagonist, and synergist.

So far from associated movements being absent in the Parkinsonian state, the reverse is the case. Not only are normally combined movements carried out (with few exceptions) just as in health; some associated movements are present which are not seen in the normal individual. The views of striatal function based on the argument that associated movements are lost in Parkinsonian states are vitiated at the outset. For example, the contention that the feet do not move when the patient rises to a standing from a sitting position is based on erroneous observation, as Wilson showed and as I can definitely substantiate.

Absence of arm-swinging in walking is the one 'associated movement' (really a movement of co-operation) which is nearly always absent (48 cases out of 50). Yet it is clear from my investigation that the movement is defective or absent largely owing to concomitant rigidity, and that in many cases the arms can as a fact be moved in walking when the attention is directed to them. But as a rule the patient does not swing the arms because it implies an effort and this effort he finds is not essential for his walking; therefore he omits it.