

EARLY DIAGNOSIS OF PARKINSON

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What a privilege it is to contribute to a Festschrift for Doctor Barraquer Ferré! It is for others to evaluate his work as a neurologist. Here I wish only to salute a man who, though seven thousand miles away I regard as one of my closest friends. His letters, which breathe such benevolent warmth, an all too short association with him in Paris in 1949 which opened to me the aristocracy of his mind and soul, cemented this friendship.

Hughlings Jackson once said "Words better our thoughts as well as define them". Many a word or term used in neurology could barely survive critical analysis and its elimination would certainly better our thoughts. Suffice it to point out the indiscriminate use of "sign", "symptom", "paresthesia", "dysesthesia"; suffice it to mention such an antiquated but still widely used term as "pseudobulbar palsy". The term "parkinsonism" is also objectionable. This term has already led to such stylistic horrors as "Rombergism". It is only hoped that "Babinskiism", "Recklinghausenism" will not follow. The suffix "ism" generally connotes a doctrine, a system, an action, an addiction, and should not be applied to a disease or a sign. In this paper the generic term "parkinson" will be used. The paralysis agitans described by Parkinson is idiopathic parkinson. Other varieties are encephalitic, toxic, infectious, traumatic, arteriosclerotic, etc.

With all natural aversion to eponyms, one must admit that the eponym "Parkinson" as well as that of "Babinski", "Wilson", has entrenched itself so deeply in the medical literature that it is difficult to see how it can ever be dislodged. It may just as well be accepted to designate the disease in general.

A fully developed parkinson of any variety is easily diagnosed. In fact, the diagnosis is often made at a glance. However, cases just beginning to show manifestations or abortive cases sometimes cause great diagnostic difficulty. The history of the patient may give no clue; in post-encephalitic parkinson it may be impossible to obtain a clear history of encephalitis or this encephalitis may have occurred as long as fifteen or twenty years previously. The insidious beginning and slow course of parkinson of any variety contributes to the difficulty of diagnosis.

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Confronted with a patient with signs of parkinson we must consider many varied morbid conditions for differential diagnosis. Among them may be mentioned: diverse degenerative and non-degenerative diseases of the brain, multiple sclerosis, exophthalmic goiter, senile tremor, familial tremor, arthritis as well as psychoneurotic, neurasthenic, hysteric states and various intoxications (alcohol, lead, mercury, etc.). Generally speaking, the diagnosis of parkinson is made much too often. One is particularly prone to misdiagnose cases of senile tremor as parkinson since both affections have many common features. Here the correct diagnosis is all important, since it affects the prognosis, completely different in the two conditions. In cases of unilateral tremor, in which form parkinson often appears, non-degenerative circumscribed lesions located outside the pallidum must be considered. Just as cases of parkinson with tremor present difficult diagnostic problems, so do those cases in which tremor is absent.

It is best to approach the early diagnosis of parkinson physiologically. What is the leading physiologic disturbance in parkinson? Universally it is accepted that the fundamental motor disturbances in parkinson can be traced to two main factors: tremor and rigidity. Which of these factors is the more important? Both to the patient and to the doctor it is often the tremor. This is understandable since tremor is the more conspicuous, the more striking manifestation of the disease. It affects the distal parts of the extremities and for that reason it is more troublesome to the patient. Many a patient with both tremor and rigidity complains only of the tremor, and disregards the rigidity. Without doubt, parkinson can and does occur when there is no tremor, as the well known term "paralysis agitans sine agitatione" indicates. On the other hand, the existence of parkinson "sine rigiditate" must be denied. I have never seen any patient with parkinson of any variety without rigidity. When a patient with parkinson has been found without rigidity, the technique in examining for tonus has undoubtedly been faulty.

In parkinson the rigidity is not only more significant than the tremor, but may precede it. There is no doubt that in parkinson, particularly in the post-encephalitic type, rigidity often comes first and the tremor later. Where does this rigidity in parkinson begin? The answer is: in the neck muscles, particularly in the flexors of the head. This leads to a sustained forward position of the head, a very early sign in parkinson. As the disease advances, this rigidity spreads, first of all to the proximal muscles of the limbs, particularly the upper, but it still remains more pronounced in the flexors of the head than in any other muscle group.

If rigidity is the fundamental sign of parkinson, if this rigidity starts first in the neck muscles, then a test for the early diagnosis of parkinson should be based on the rigidity of these neck muscles. From one of my teachers — it might have been Cassirer — I learned to test every patient with parkinson for rigidity of the neck muscles. The palm of the examiner is placed on the top of the patient's head; the patient is made to relax,

the head is moved passively in every direction, the resistance is appraised. This is, of course, a crude method to test for rigidity of the neck muscles. The following test is much more accurate.

HEAD DROPPING TEST

The technique is as follows: The patient lies supine on the examining table without a pillow. The surface should be padded, not hard. As the patient often has a tendency to raise his head, he should be made to relax completely; his attention is diverted and he is engaged in conversation. His head rests on the table surface. The examiner places his left hand, dorsum on the table, under the patient's occiput, which then lies in the palm of the examiner. With his right hand, the examiner suddenly, unexpectedly, and briskly lifts the patient's head and lets it drop. In normal persons the head drops back with force, like dead weight. It is as if there were no tonus of the neck muscles, as if the head were attached to the body by a string. The fall is so heavy that were it to occur on a hard surface, the patient could sustain an injury. This complete lack of tonus of the neck muscles is in gross contradistinction with the marked tonus they exhibit while the patient is erect. One is amazed at the sharp impact with which the head falls, heavily and inertly. The examiner can both see and feel the impact of the falling head. It is advisable to repeat the test. If, after such repetition varying results are obtained, the heaviest drop counts. In all others, it must be assumed that some active interference on the part of the patient has taken place. The essential prerequisite for this test is complete relaxation in the patient. When in rare instances this relaxation cannot be achieved, the test must be given up for the time being.

The test can also be performed with the patient lying prone, or on either side, but it is much more easily performed and the results are less difficult to evaluate when the patient lies supine. In parkinson, the rigidity of the neck muscles is greater in the flexors than in the retractors of the head. The head dropping test is therefore more delicate when the patient lies supine than when he lies prone.

The most striking change from the normal in this test is seen in parkinson, even in its earliest stages. Here the head drops slowly and gently; the downward movement is, so to speak, reluctant, hesitant, lazy. This slow dropping of the head shows that, though the rigidity of the muscles in parkinson is diminished when the patient lies in a horizontal position, it still remains considerable. The movement is of the same character as that felt by the examiner when he passively moves a rigid extremity. The downward movement of the head maintains an even flow, is smooth, uniform from beginning to end. There is no sudden "give", no erratic unevenness. If the patient interferes voluntarily and makes a movement with his head, this can be distinguished immediately by the examiner.

The examiner can appraise the character of the motion not only by sight but by touch as well. He can feel the impact of the dropping head on the palm of his hand. Compared with the violent impact of the normal, the head in parkinson hits the palm of the examiner softly and smoothly. The difference is unmistakable, even in the earliest stages of parkinson. Patients with parkinson who have never complained of rigidity may show a head dropping test typical of this disease.

In advanced cases the test is superfluous. Here the rigidity of the neck muscles is too obvious to need any special test. This rigidity is drastically illustrated by the fact that the patient with advanced parkinson, lying supine, keeps his head bent and off the surface indefinitely. The position is, so to speak, frozen.

In parkinson of any variety the head dropping test has been found abnormal so regularly and in such early stages, and this abnormality has been so clear and unequivocal that it can be stated: *if the head dropping test is normal, parkinson can be excluded.*

This test is of particular importance in differential diagnosis: parkinson versus senile tremor. Cases of tremor have been seen, diagnosed as parkinson by neurological authorities. Later the head dropping test was found to be normal. This finding led to further clinical investigations which proved beyond a doubt that the patient suffered from senile tremor and not from parkinson. The differential diagnosis between these two conditions may be very difficult, the tremor *per se* may not give any decisive diagnostic clue for differential diagnosis. The outstanding characteristic of senile tremor, tremor of the head, may also occur in parkinson. Many authors even state that senile tremor is just a "forme fruste" of parkinson. However, a differential diagnosis is of importance particularly with regard to prognosis. since senile tremor is a benign, non-progressive lesion, while parkinson is not. In this differential diagnosis — parkinson tremor versus senile tremor — the head dropping test is of decisive importance. The cardinal fact is that there is no neck rigidity in senile tremor and the head dropping test here is always unmistakably negative. This decides the issue.

The head dropping test, as found in parkinson, is found also in other extrapyramidal affections which show rigidity of the neck muscles such as progressive lenticular degeneration, torsion dystonia, striatal torticollis, pallidal degeneration. Generally speaking, an abnormal head dropping test in any diffuse morbid condition of the brain indicates concomitant involvement of the extrapyramidal system. In pyramidal affections even when they are bilateral, remarkably enough, the head dropping test deviates but little from the norm. Changes seen here are very slight and can be distinguished easily from those seen in affections of the extrapyramidal system.

TEST FOR PENDULOUSNESS OF THE ARM IN WALKING

This test is so well known that little need be said about it. In parkinson rigidity affects the neck muscles first and most of all, and then the proximal muscles of the upper extremities. This very rigidity reduces the automatic pendulousness of the arms in walking. The diminution of pendulousness is particularly conspicuous in unilateral parkinson. It may decide a doubtful diagnosis.

To examine for pendulousness of the arms, the patient should walk in a more spacious place than the usual examining room, for instance in a hall or in any open space. Heavy outer garments should be removed. The patient should hold nothing in his hands. He should be asked to walk at varying speeds. Since in this examination the excursion of the arm is all important, it can be more easily appraised if the examiner, keeping pace with the patient, walks abreast of him and watches him from the side. The degree of pendulousness is best judged by noting the forward excursion of the arm, i. e., how far the hand moves in front of the trunk. Every person has his characteristic pendulousness of the arms in walking. It varies both in character and in range from person to person. It is of no particular importance if the excursion is short on both sides, unless there are other definite signs of parkinson. More important is the difference in range between right and left. In parkinson, pendulousness of the arm on one side, particularly the forward excursion, may be diminished very early in the course of the disease. For some time this may even go unnoticed by the patient. A peripheral or a pyramidal lesion affecting the side of the diminished range can easily be excluded. No other disease of the nervous system reduces the pendulousness so early and so obviously as does parkinson. If some anomaly is found, the examination should be repeated or extended. It is surprising how often at first one finds changes in pendulousness of the arms which later cannot be confirmed. Only deviations found on repeated examinations are of value, but this value is fundamental.

SHOULDER SHAKING TEST

The examiner, facing the standing patient, places his hands on the patient's shoulders and moves them in brisk jerks and with varying speed and force alternately to and fro. Swinging of the patient's arms results. In unilateral parkinson this swinging is diminished. By this test it is possible, to some degree, to appraise the diminution quantitatively. The examiner, keeping his movement equal on both sides, gradually and slowly reduces the force and range of his jerks. He thus causes a reduced shoulder motion, and consequently reduced pendulousness. If the pendulousness of one arm is diminished due to parkinson, the examiner reaches a point when only one arm swings, while the other remains static. The arm that still swings is normal or less affected.

Another technique of shoulder shaking which sometimes gives even better results than the one just described is as follows: The patient keeps his feet firmly on the ground; the examiner, his hands on the patient's shoulders, gently rocks the patient's trunk back and forth. Here the movement is not alternate, right and left, but the shoulders are moved in the same direction back and forth. This produces a simultaneous swinging of both arms. The one that describes the greatest excursion is the healthy arm or the one less affected.

The shoulder shaking test is useful to confirm the findings of pendulousness of the arms in walking or to help in making the diagnostic decision in doubtful cases.

This test may be valuable in still another way. Not only extrapyramidal rigidity produces diminished pendulousness of the arm in walking, but a homolateral cerebellar affection may do the same. In this case, though there is no hypertonia as in extrapyramidal disease, but cerebellar hypotonia instead, still the affected arm swings less. The pathophysiology is not the same in the two conditions. Extrapyrarnidal affections diminish pendulousness through rigidity of the affected muscles. Cerebellar affections produce diminution of pendulousness through "decompensation of movement". In the latter case, the arm though hypotonic, does not move synchronously with the contralateral leg in walking, due to cerebellar dys-synergia, i.e., the lack of synergic movements of different parts of the body. If a patient shows diminution of the pendulousness of one arm in walking, the question arises as to whether this is extrapyramidal or cerebellar in origin. The shoulder shaking test will decide the issue. If, on shoulder shaking, the affected arm moves less, the affection is extrapyramidal; if it moves, or it moves more, cerebellar.

ARM DROPPING TEST

This test, too, serves to help diagnose and qualitatively appraise rigidity of the shoulder muscles in parkinson. With both arms hanging loosely at his sides, the patient stands before the examiner. He is made to relax his arms as far as possible. The examiner stretches out his own arms, and slides his hands between the patient's pelvis and arms, so that the dorsum touches the inner surface of the patient's wrist. The attention of the patient is diverted. Suddenly and unexpectedly the examiner, with a quick, forceful movement of both his hands simultaneously, throws the patient's arms upward and allows them to drop, but meanwhile quickly brings his own hands back to their former position at the patient's pelvis. His hands receive the impact of the patient's arms as they fall. Thus the examiner can see, feel, and hear the differences in the fall of the arms. In unilateral parkinson, the fall of the arm is retarded on the affected side. It hits the examiner's hand less heavily, less inertly, and the sound is duller than that on the normal side.

A similar technique can be used to appraise the tonus in the forearm. The patient sits relaxed with his forearms on his thighs. The examiner sits in front of the patient and with sudden, jerky, tossing movements, flings the forearms upward from the wrists. The rigidity of the muscles of the forearm documents itself in the diminution of the range of the upward movement and the retardation of the dropping movement.

TEST FOR PENDULOUSNESS OF THE LEGS

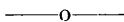
This simple and very reliable test serves quickly to appraise the muscle tonus of the lower extremities. The patient sits on the examining table, his legs hanging freely over the edge. He is made to relax. The examiner lifts the patient's legs and allows them to drop, observing the length of time they swing. The extrapyramidal rigidity very markedly shortens this swinging time. In unilateral parkinson, the difference in swinging time between the affected and the healthy leg is very striking. The following observation has been repeatedly made: a patient in the earliest stage of parkinson shows a slight diminution of pendulousness of one arm on walking and also when the shoulder shaking test is performed. He shows no other sign nor symptom. On specific interview, he denies that the homolateral leg is in any way affected. His gait is normal and yet, when the test for pendulousness of the legs is performed, the pendulousness of the homolateral leg is markedly diminished. There is no doubt that the *diminution of the pendulousness of one leg may indicate involvement of this leg in parkinson before any other sign manifests itself.*

EXAGGERATION OF THE ORBICULARIS OCULI REFLEX

This reflex is a deep muscle reflex of the orbicularis oculi muscle. Concussion and stretching of this muscle result in closure of the eyes, in winking. The orbicularis oculi muscle reacts also with contraction to many other stimuli, visual, auditory, acoustic, but these reflexes are cortical. The stimuli evoking these reflexes, particularly a visual stimulus, must be maximally avoided when the deep muscle reflex is elicited. To elicit this reflex it is best to tap the glabella gently with a reflex hammer and to avoid startling the patient. Since the muscle lies directly on the bone, and the threshold of its reflex contraction is very low, the mildest tapping, by producing concussion of the bone, produces concussion and stretching of the muscles and elicits a reflex contraction of the muscles and blinking. This reflex is markedly exaggerated in parkinson of any variety. This is found so regularly that it can serve in differential diagnosis. It is remarkable that in pyramidal bilateral tract involvement, in supranuclear bulbar palsy this exaggeration is not found. It is striking to see the exaggeration of this reflex in parkinson, which is in sharp contrast with the infrequent blinking found in that disease.

CONVERGENCE PARESIS OF THE EYES

On attempted convergence the eyes do not move inward, or they do so insufficiently. The paresis may be asymmetrical. It may appear very early in parkinson, particularly in the post-encephalitic form.



These tests are the most reliable for early diagnosis of parkinson. There are many other tests which help in diagnosis, but these give unmistakable results only in pronounced cases, and may serve more to confirm the diagnosis than to establish it. Among these tests the following may be mentioned briefly: the hand and fingers, particularly the thumb, assume an abnormal position; fine finger movements show easy exhaustibility; when the forearm is flexed toward the upper arm, every movement is accompanied by tightening of the biceps muscle; when the patient, with his eyes closed, stretches his arms out horizontally in front of him, the arms have a tendency to deviate inward and downward; the patient has a tendency to write small characters, or to decrease their size as he continues to write (micrographia).

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