How is a New Natural History of Tremblings, Agitations, and Shaking Created in 1817?

Brian Hurwitz 7th Narrative and Health Behavioural Science Meeting, London Sept 2014

ESSAY

ON THE

SHAKING PALSY.

BY

JAMES PARKINSON, MEMBER OF THE ROYAL COLLEGE OF SURGEONS.

LONDON: PRINTED BY WHITTINGHAM AND ROWLAND. Commel Street,

FOR SHERWOOD, NEELY, AND JONES, PATERNOSTER ROW.

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Parkinson's disease: Diagnosis and management in primary & secondary care

CG35 June 2006



Parkinson's disease is a progressive neurodegenerative condition resulting from the death of the dopamine-containing cells of the substantia nigra. There is no consistently reliable test that can distinguish it from other conditions that have similar clinical presentations. The diagnosis is primarily clinical, based on a history and examination.



People with condition present with the symptoms and signs of **parkinsonism**

-bradykinesia -rigidity

-rest tremor.

Predominantly a movement disorder, but other impairments frequently develop including depression and dementia. Autonomic nervous disturbances and pain (which can be a presenting feature) may later ensue, and the condition progresses to cause significant disability and handicap with impaired quality of life for the affected person. Family and carers may also be affected indirectly.



UK PARKINSON'S DISEASE SOCIETY BRAIN BANK CLINICAL DIAGNOSTIC CRITERIA* 3 steps

Step 1. Diagnosis of Parkinsonian Syndrome

- Bradykinesia
- At least one of the following
- 1 Muscular rigidity
- 2 4-6 Hz rest tremor
- 3 postural instability **not** caused by primary visual, vestibular, cerebellar, or proprioceptive dysfunction



Step 2 Exclusion criteria for Parkinson's

 history of repeated strokes with stepwise progression of parkinsonian features

- history of repeated head injury
- history of definite encephalitis
- neuroleptic treatment at onset
- sustained remission
- strictly unilateral features after 3 years
- supranuclear gaze palsy
- cerebellar signs
- early severe autonomic involvement
- •Babinski sign +ve



Step 3 supportive prospective positive criteria for Parkinson's disease

Three or more required for diagnosis of definite Parkinson's disease in combination with step one

- Unilateral onset
- Rest tremor present
- Progressive disorder
- Persistent asymmetry affecting side of onset most
- Excellent response (70-100%) to levodopa
- Severe levodopa-induced chorea
- Levodopa response for 5 years or more
- Clinical course of ten years or more.

Hughes AJ, Daniel SE, Kilford L, Lees AJ. Accuracy of clinical diagnosis of idiopathic Parkinson's disease. A clinico-pathological study of 100 cases. JNNP 1992;55:181-184.

Parkinson's is a <u>progressive neurological condition</u>. People with Parkinson's don't have enough of the chemical dopamine because some nerve cells in their brain have died.

Every hour, someone in the UK is told they have Parkinson's.

Without dopamine people can find that their movements become slower so it takes longer to do things. The loss of nerve cells in the brain causes the symptoms ...

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Main symptoms of Parkinson's <u>Tremor (shaking)</u> <u>Slowness of movement</u> <u>Rigidity (stiffness)</u>

Physical and other symptoms of Parkinson's

Bladder and bowel problems Eye problems Falls and dizziness Falls and dizziness Fatigue Freezing Pain Restless legs syndrome Skin, scalp and sweating problems Sleep problems Speech and communication problems Swallowing problems

Mental health and Parkinson's <u>Anxiety</u> <u>Dementia</u> <u>Depression</u> <u>Hallucinations and delusions</u> <u>Memory problems</u>

Help and support

Parkinson's diagnosis

It's not easy to diagnose Parkinson's. There are no laboratory tests so it's important that the diagnosis is made by a specialist.



Summary

Parkinson's disease is a common, progressive, bradykinetic disorder that can be accurately diagnosed. It is characterised by the presence of severe pars-compacta nigral-cell loss, and accumulation of aggregated α -synuclein in brain stem, spinal cord, and cortical regions. The main risk factor is age.

Susceptibility genes including α -synuclein, leucine rich repeat kinase 2 (*LRRK-2*), and glucocerebrosidase (*GBA*) have shown that genetic predisposition is another important causal factor. Dopamine replacement therapy considerably reduces motor handicap, and effective treatment of associated depression, pain, constipation, and nocturnal difficulties can improve quality of life. Embryonic stem cells and gene therapy are promising research therapeutic approaches.



2055–2066

Parkinson's disease commonly presents with impairment of dexterity or, less commonly, with a slight dragging of one foot.

The onset is gradual and the earliest symptoms might be unnoticed or misinterpreted for a long time. Fatigue and stiffness are common but non-specific complaints. Work colleagues or family members might notice:

- a stiff face, a hangdog appearance
- flexion of one arm with lack of swing
- a monotonous quality to the speech
- and an extreme slowing down.



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- a stiff face, a hangdog appearance 'downcast', miserable, wretched, shame-faced, 'sheepish': selfconscious; tail-between-legs appearance
- flexion of one arm with lack of swing
- a monotonous quality to the speech
- and an extreme slowing down.



Lees A, Hardy J, Revesz T. Parkinson's disease. *Lancet* 373:13–19 June 2009, 2055–2066

These changes are rarely noticed by the patient. The early physical signs are often incorrectly put down to old age, misery, introspection, or rheumatism. A lag of 2–3 years from the first symptoms to diagnosis is not unusual.

Once the diagnosis has been confirmed, patients and their families often start to remember potentially relevant symptoms and signs going back more than a <u>decade</u>. Early difficulties with coordination might have been blamed on faulty equipment, such as a keyboard that keeps typing a letter twice; a towel that does not dry properly; or a self-winding watch that keeps stopping despite reassurances from the manufacturer. Each neurologist has his anecdotes about unusual clinical presentations.



Lees A, Hardy J, Revesz T. Parkinson's disease. *Lancet* 373:13–19 June 2009, 2055–2066

Kinnier Wilson described the case of a colleague who remained abnormally still in his seat during a medical conference, and another of a friend who commented that his first symptom was that he could walk more easily on a pebbled beach than in a crowded street.

Loss of sense of rhythm and a tendency to swim in circles are two personal recollections. Early motor symptoms can be subtle and easily missed. A change in a patient's writing can be present for several years before diagnosis, with a tendency to slope usually in an upward direction and for the writing to get progressively smaller and more cramped after a line or two.



Early loss of smell is occasionally spontaneously reported but many patients are unaware of hyposmia until they are formally tested. Disturbed sleep—including shouting out, flailing movements of arms and legs, and falling off the bed during dreaming—might only be noticed if the patient's spouse is questioned.

Complaints of falls (especially backwards), fainting, urinary incontinence, prominent speech, disturbed swallowing, amnesia, or delirium should raise the possibility of an alternative diagnosis.



Lees A, Hardy J, Revesz T. Parkinson's disease. *Lancet* 373:13–19 June 2009, 2055–2066

In the late stages of Parkinson's disease, the face of patients is masked and expressionless, the speech is monotonous, festinant, and slightly slurred, and posture is flexed simian with a severe pill rolling tremor of the hands.

Freezing of gait for several seconds can happen when attempting to enter the consulting room and, when starting to move again, the patient tends to move all in one piece with a rapid propulsive shuffle. These motor blocks lead to falls. All dextrous movements are done slowly and awkwardly, and assistance is needed for dressing, feeding, bathing, getting out of chairs, and turning in bed. Constipation, chewing and swallowing An Essay on the Shaking Palsy **CONTENTS**

- I. Definition—History—6 Illustrative cases
- ii. Pathognomonic symptoms examined Tremor coactus Scelotyrbe festinans
- iii. Shaking palsy distinguished from other diseases with which it may be confounded
- iv. Causes proximate & remote—3 Illustrative cases
- v. Considerations respecting the means of cure.

An Essay on the Shaking Palsy
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v. Considerations respecting the means of cure.

Shaking Palsy (Paralysis Agitans)

Involuntary tremulous motion, with lessened voluntary power, in parts not in action, and even when supported; with a propensity to bend the trunk forwards, and to pass from a walking to a running pace: the senses and intellects being uninjured.

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The disease is of long duration: to connect the symptoms which occur in its later stages with those which mark its commencement, requires a continuance of observation of the same patient ... even for years.

HISTORY

So slight and nearly imperceptible are the first inroads of this malady, and so extremely slow its progress, that it rarely happens, that the patient can form any recollection of the precise period of its commencement.

The first symptoms... are a slight sense of weakness, with a proneness to trembling in some particular part; sometimes in the head, but most commonly in one of the hands and arms.

[A]s the disease proceeds, the hand fails to answer with exactness to the dictates of the will. The legs are not raised to that height, or promptitude which the will directs... [W]riting can now be hardly at all accomplished; and reading, from the tremulous motion, is accomplished with some difficulty. Whilst at meals the fork not being duly directed frequently fails to raise the morsel from the plate which, when seized, is with much difficulty conveyed to the mouth.

Commencing ... in one arm, the wearisome agitation is borne until beyond sufferance, when by suddenly changing the posture it is for a time stopped ... to commence, generally in less than a minute in one of the legs Harassed by this tormenting round, the patient has recourse to walking, a mode of exercise to which sufferers from this malady are in general partial....

As the malady proceeds ... the propensity to lean forward becomes invincible, and the patient is thereby forced to step on the toes and fore part of the feet, whilst the upper part of the body is thrown so far forward as to render it difficult to avoid falling In some cases... the patient is ... irresistibly impelled to take much quicker and shorter steps, and thereby to adopt unwillingly a running pace. Harassed by this tormenting round, the patient has recourse to walking, a mode of exercise to which sufferers from this malady are in general partial....

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The saliva fails of being directed to the back part of the fauces, and hence is continually draining from the mouth, mixed with the particles of food... As the debility increases the tremulous agitation seldom leaves him; but even when exhausted nature seizes a small portion of sleep, the motion becomes so violent as not only to shake the bedhangings, but even the floor and sashes of the room.

The chin is now almost immovably bent down upon the sternum. The slops with which he is attempted to be fed, with the saliva, are continually trickling from the mouth. The power of articulation is lost. The urine and faeces are passed involuntarily; and at the last, constant sleepiness, with slight delirium, and other marks of extreme exhaustion announce the wished-for release.

CASE 1

Almost every circumstance noted in the preceding description, was observed in a case which occurred several years back... The subject of this case was a man rather more than fifty years of age, who had industriously followed the business of a gardener, leading a life of remarkable temperance and sobriety.

The commencement of the malady was first manifested by a slight trembling of the left hand and arm... In this case, every circumstance occurred which has been mentioned in the preceding history.

ABOUT CASE 1

It occurred several years back, and ... from the particular symptoms which manifested themselves in its progress; from the little knowledge of its nature acknowledged to be possessed by the physician who attended; and from the mode of its termination; excited an eager wish to acquire... further knowledge of its nature.

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<u>Case 5</u>

...a hand on each shoulder, until, by gently swaying backward and forward, the patient had placed himself in equipoise; when, giving the word, he would start in a running pace, the attendant sliding from before him and running forward, being ready to receive him and prevent his falling.





Entrance of TOTTENHAM COURT ROAD TURNPIKE, with a View of J. TAMES'S CHAPEL

London Rublillid Mar 1" 1813, at Ackermann's Gallery, No in Strand









<u>CASE 6</u>

It being asked, if whilst walking he felt much apprehension from the difficulty of raising his feet if he saw a rising pebble in his path? He avowed, in a strong manner, his alarm on such occasions; and it was observed by his wife, that she believed, that in walking across the room, he would consider a difficulty the having to step over a pin.

<u>CASE 6</u>

... he being then just come in from a walk, with every limb shaking, threw himself rather violently into a chair, and said, "Now I am as well as ever I was in my life". The shaking completely stopped; but returned within two minutes....

<u>CASE 6</u>

It... being asked if he walked under much apprehension of falling forwards? he said he suffered much from it; and replied in the affirmative to the question, whether he experienced any difficulty in restraining himself from getting into a running pace. It being asked, if whilst walking he felt much apprehension from the difficulty of raising his feet, if he saw a rising pebble in his path?

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In conclusion

Elements:

Indistinct beginning - unnoticed by patient

Long duration condition

Affects older people (men)

In conclusion

Worsening trajectory involving multiple body systems

Clear temporal shape.

In conclusion

Specific features

- Tremor, shaking, agitations, tremulousness 'nervous body'
- Shaking when limb 'supported', 'when unemployed' 'tremor at rest'
- V specific posture and gait disorder
- Swallowing difficulty saliva and tongue problems
- Difficulties reading, writing, walking and eating

Subjective experience – Family/carer involvement