Fast Facts: Parkinson’s Disease

Fourth edition

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Declaration of Independence
This book is as balanced and as practical as we can make it.
Ideas for improvement are always welcome: feedback@fastfacts.com

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Introduction

Parkinson’s disease is one of the most important disabling diseases of later life. It was first described by James Parkinson in 1817 in *An Essay on the Shaking Palsy*. Since then, the disease has become the pathfinder for other neurodegenerative disorders, starting with the discovery of dopamine deficiency within the basal ganglia, which led to the development of the first effective treatment for a progressive neurodegenerative condition.

Dopamine-replacement therapy substantially reduces the motor symptoms of Parkinson’s disease in most patients, improving their quality of life and initially appearing to decrease mortality. In recent times, however, the non-motor symptoms of Parkinson’s disease – the long-neglected Cinderella of Parkinson’s management – have emerged as the key determinant of quality of life and challenge to treatment. Depression, sleep dysfunction, fatigue, pain and anxiety have been identified as some of the key symptoms of the illness, while visual hallucinations, dementia and falls can result in hospitalization and institutionalization. In addition, Parkinson’s disease is associated with considerable caregiver stress.

Given the burdens that Parkinson’s disease can impose, this book has been designed to provide doctors, nurses and therapists with the latest information in order to improve as much as possible the lives of patients with Parkinson’s disease and related disorders. While most books focus on drug therapy, genetic research or treatment for motor symptoms, here we focus on the ‘holistic’ care of patients.

Since earlier editions there have been many advances in the diagnosis and management of Parkinson’s disease and in the care available for individuals with the condition. New genes have been described, and new methods to aid diagnosis such as transcranial ultrasound have been developed. Meanwhile new clinical trials are now being reported with non-motor outcomes.

Developments in therapy such as human-fetal-cell transplantation or gene therapy/stem cell-based therapy continue to be assessed. Meanwhile, the therapeutic armamentarium continues to expand, with
improved injection devices for apomorphine, intrajejunal infusion of levodopa and refinement of delivery of deep brain stimulation.

Non-motor symptoms are tied to progression of underlying disease, and studies have tried to address interventions that slow disease progression. Encouraging but inconclusive results have emerged using the monoamine oxidase B (MAOB) inhibitor rasagiline in a delayed-start design, while other trials such as the PROUD study have failed to produce significant results.

Animal models underpin the key developments in understanding pathogenesis and treatment of neurodegenerative conditions, and research continues to seek a true animal model of Parkinson’s disease, one that shows progressive neurodegeneration with Lewy body formation and motor as well as non-motor symptoms.

An overwhelming body of evidence and patient testimony underlines the importance of non-motor symptoms, the need for multidisciplinary care and the use of tools that empower patients. Guidelines highlighting the important role of multidisciplinary care have been published, while the focus of research has shifted from a bias towards motor symptoms to non-motor symptoms, with the publication of specific tools to assess and flag these important problems.

In the updating of this fourth edition of *Fast Facts: Parkinson’s Disease*, we have sought to address all of these aspects. Importantly, we have consulted with patients and sought to reflect their perspectives throughout. People with Parkinson’s disease require multidisciplinary professional care. When they ask questions they must feel assured that the answers are well informed and correct. This truly useful and unique resource is therefore essential reading for the whole team.

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The Parkinson’s journey

Prodromal Parkinson’s disease
The Parkinson’s journey may begin long before diagnosis, as Parkinson’s disease is now recognized to have a prodromal period dominated by a number of non-motor symptoms. These are late-onset hyposmia or anosmia, rapid eye movement (REM) behavior disorder, episodes of major depression or anxiety, and excessive daytime sleepiness. This prodromal period could last up to 20 years before awareness of the motor symptoms that mark the initial or ‘stable’ period of the condition. Those who have the non-motor prodrome will have had unexplained symptoms for years, while those with a dominant motor presentation may have had symptoms for only a few months.

Diagnosis
For the clinician, diagnosis is based on clinical presentation, as there is still no reliable diagnostic test. While the cardinal motor symptoms
and signs of rest tremor, akinesia and rigidity remain the mainstay of diagnosis, the prodrome of non-motor symptoms described above develops in the majority of patients. At least half of all patients experience mood disturbance with anxiety or depression at some stage of the illness, including in the prodromal phase.

“It was almost a relief to be diagnosed with PD. Up until then I had been told I was suffering from work-related stress and anxiety. I thought it was PD before it was diagnosed so it did not come as a shock. In some ways it was good to know that there was a physical reason why I could no longer cope at work.”

**Delivering the diagnosis.** Care must be taken in how the diagnosis is delivered. Patients are often fearful and may come to the clinician with preconceptions based on information overload from the internet, or a limited understanding or experience of the disease from the media or a relative. It is best to break the news of a patient’s diagnosis in the presence of their spouse, partner or other family members.

Often, little information is retained from that first consultation and it is necessary to meet again within 2–3 weeks when the patient’s initial shock has subsided.

“When my husband finally emerged from that consultation, his face was completely expressionless. He rushed past me, down the stairs, over the parking lot and I ran after him ... there he sat with his head in his hands and tears streaming down his face.”

“Please remember the effects of the diagnosis on the spouse as well. The shock can be just as bad for them and the long-term effects can be severe, as they have to support their loved one as they watch what they go through.”
When, in 1817, James Parkinson first described the features of paralysis agitans in 6 patients, he did not refer to the typical cogwheel rigidity, and mistook bradykinesia for paralysis. Nevertheless, his description of the tremor, posture (Figure 3.1) and clinical course of the disease has stood the test of time and remains valid today. Most cases of Parkinson’s disease are easily recognizable at an early stage, but many are missed if tremor is absent; gradual slowing in performance may be instead attributed to aging or aches and pains, and loss of function may be ascribed to other causes.

Figure 3.1 Paralysis agitans, as first described by James Parkinson, depicting the characteristic fixed posture. Neurologist Sir William Richard Gowers drew this illustration in 1886 as part of his documentation of Parkinson’s disease in *A Manual of Diseases of the Nervous System*. 
Parkinsonism is a clinical syndrome (Table 3.1) and may have a number of causes. When the condition appears to be idiopathic and, in particular, responds to levodopa therapy, it is referred to as Parkinson’s disease.

**Early motor and non-motor indicators**
Signs may be subtle in the early stages of the disease. In suspected cases it is often helpful to ask patients what tasks they find difficult.

### TABLE 3.1
Features that support a diagnosis of parkinsonism

**Essential features**
- Bradykinesia and one (or more) of the following:
  - tremor (resting)
  - rigidity (cogwheel or lead-pipe; see page 46)
  - postural instability

**Additional motor features**
- Fixed, stooped posture
- Dystonic postures, e.g. striatal hand, striatal toe
- Hypomimia (‘masked’ face)
- Shuffling, short-step gait (with or without festination)
- Freezing episodes (sometimes known as paradoxical akinesia)

**Additional non-motor features**
- Late-onset hyposmia
- Depression and anxiety
- Constipation
- Bladder symptoms
- Pain (usually unilateral on the affected side)
- Subtle mental and cognitive disturbance (mild cognitive impairment)