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# Time to retire the term "atypical Parkinsonism"

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A B S T R A C T

Variable and imprecise use of the term "Parkinsonism" has produced confusion among patients, clinicians, caregivers, and researchers. The term "atypical Parkinsonism" can be dispiriting and even demeaning to patients. We therefore propose a slight terminological revision for Parkinsonian disorders to eliminate the terms "typical" and "atypical" and to confine the term "Parkinsonism" to the phenotypic syndrome and not to disease entities. We also suggest replacing the "atypical Parkinsonism" term with "Parkinson-like disorders" to emphasize that it is not PD but does resemble it at some level. "Parkinson-like disorders" would comprise the primary neurodegenerative Parkinsonian disorders other than PD, chiefly MSA, DLB, PSP, and CBD, and would be consistent with the existing neurologic terms, "Huntington's disease-like," "polio-like," and "stroke-like." We would continue the practice of referring to the non-degenerative Parkinsonian disorders as "secondary Parkinsonian disorders."

## 1. Background

Three major nosologic entities are characterized by Parkinsonism: Parkinson's disease (PD; a degenerative disorder characterized by Lewy bodies and emphasizing the brainstem), atypical Parkinsonian disorders (degenerative disorders with a phenotype resembling that of PD but either without prominent Lewy bodies or without brainstem emphasis), and secondary Parkinsonian disorders (typically with fragments of the PD phenotype and typically of non-degenerative cause). We propose new terms that may be more logical, consistent, and compassionate.

Parkinsonism is a syndrome of bradykinesia with rigidity or tremor or both. It is not a disease but rather a constellation of clinical features occurring in a variety of neurological diseases, most but not all of which are neurodegenerative. As a group of physical findings, "Parkinsonism" is an inherently a singular noun. However, the term is commonly applied, including in a plural form, to any disorder featuring that group of findings.

James Parkinson named the disorder "the shaking palsy" in his seminal 1817 description, although the syndrome had been precisely described much earlier [1–3]. The term "paralysis agitans" was also used over the decades without adding clarity or information. In 1872, Charcot renamed the condition "Parkinson's disease" both on honorific grounds and because he found that true weakness ("palsy") was usually absent. The precise definition of PD has evolved since then [4–6].

The origin of the term "Parkinsonism" is more difficult to date. It does not appear in the 14-page discussion of "paralysis agitans" in the 1891 edition of the leading American neurology textbook of the time [7]. However, the term was in use by 1926 in the context of post-encephalitic Parkinsonism [8].

## 2. Logical issues with current terminology

If we agree that "Parkinsonism" is a clinical phenotype, then using it also to apply to specific disease entities only creates confusion. By that logic, the plural form, "Parkinsonisms," makes no sense.

In applying the term "atypical Parkinsonism" to disorders such as PSP and MSA, neurologists understand that they are not merely unusual variants of PD, but fundamentally different disease entities, albeit with some pathoanatomic overlap with it. Aside from this logical and scientific inconsistency, the term "atypical" conveys an "other-ness" or a hopelessness born of insufficient scientific understanding.

The term "Parkinson-plus" has also been applied to non-PD disorders featuring a Parkinsonian phenotype. It refers to the prominence of such additional features as dysautonomia or ataxia in MSA, gaze palsy and early falls in PSP, and cognitive loss and fluctuating encephalopathy in DLB. But these disorders could as well be considered "Parkinson-minus" in that rest tremor and levodopa response are typically rare, minor, or absent. Besides, the term "-plus" implies that those disorders are supersets of PD when, as noted, they merely share some features and are fundamentally different diseases at the pathogenetic level.

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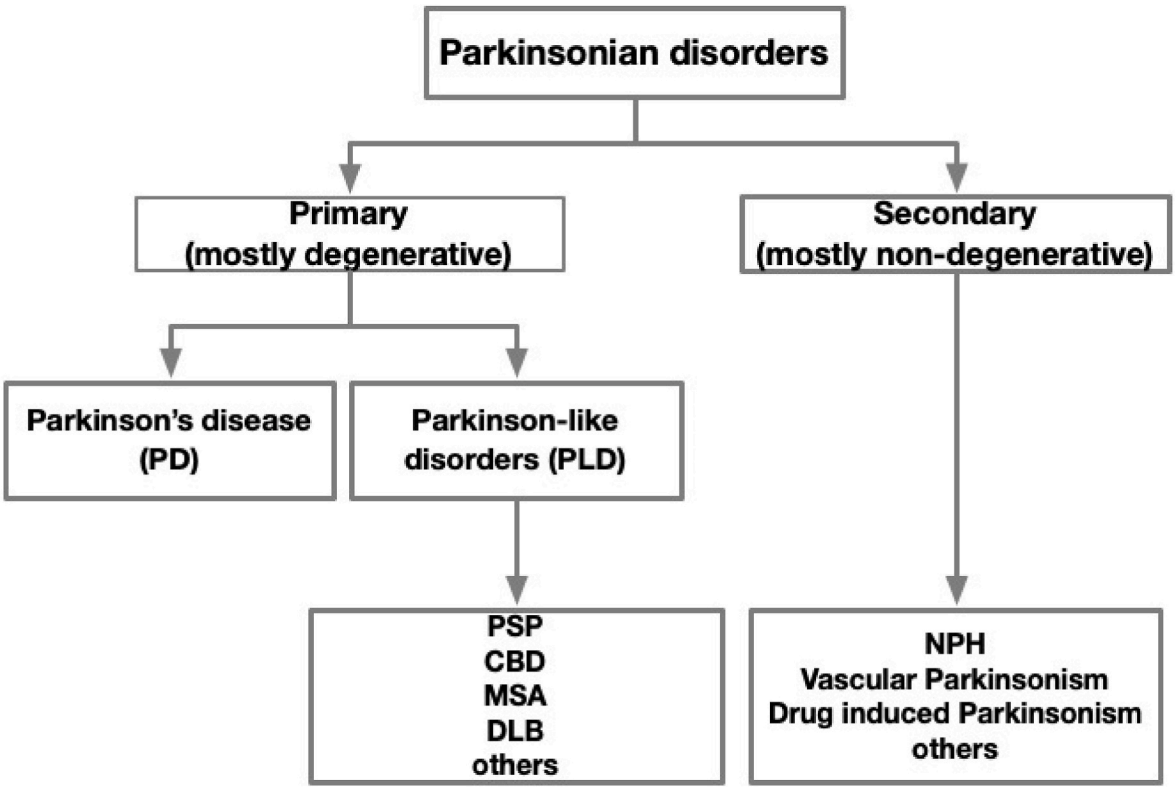


**Fig. 1.** Proposed revised nosology of the Parkinsonian disorders.

## 3. Some solutions

### 3.1. Secondary Parkinsonian disorders

Disorders with well-understood etiology and/or pathogenesis may display a minor or unusual (i.e., "secondary") Parkinsonian motor component. Chief among these are such common conditions as normal pressure hydrocephalus, vascular Parkinsonism, and drug-induced Parkinsonism. For all these, a poorly understood neurodegenerative component has been suspected, but only as a minor contributor. The category of secondary Parkinsonian disorders also includes dozens of rare conditions with known etiology, such as Wilson's disease, anti-IgLON5 encephalopathy, and Niemann-Pick disease type C.

### 3.2. Primary Parkinsonian disorders

If we recognize "secondary" Parkinsonian disorders, then it is sensible to continue using the term "primary" for PD itself and for other Parkinsonian disorders where Parkinsonism is the first or main feature, at least in a large majority of cases. Most primary Parkinsonian disorders are neurodegenerative, but advances in etiologic understanding are starting to move many of these entities out of the "degenerative" rubric, a process that will only accelerate.

### 3.3. Parkinson's disease

We suggest no change to current usage of this term. When Parkinsonism is gradually progressive with no red flags to suggest a non-PD condition and there is a good response to levodopa, a diagnosis of PD as its cause is likely and can be confirmed using formal, published diagnostic criteria [9]. In this situation, stating that the patient has Parkinsonism caused by PD is logical and appropriate. We should not require that PD be neurodegenerative (i.e., progressive with cell loss and no clear etiology) as our recognition of specific genetic and environmental etiologies of PD grows.

### 3.4. A new term: "Parkinson-like disorder"

We propose the term "Parkinson-like disorder" for the degenerative disorders presently termed the "Parkinson plus" or "atypical Parkinsonian" disorders. Chief among these are PSP, MSA, CBS, and DLB but the term also includes frontotemporal dementia with Parkinsonism, the striato-pallidal degenerations, postencephalitic Parkinsonism, spinocerebellar ataxia type 3 (and other SCAs with Parkinsonism), and juvenile-onset Huntington's disease. Clearly, many of these conditions have well-understood genetic, immune, or infectious etiologies and are not considered "degenerative." The "-like" construction has ample neurological precedent in "Huntington-like disorders" "polio-like syndromes," and "stroke-like episodes." Most of the diseases in this category are more rapidly progressive than PD and are less treatable, while others, such as vascular Parkinsonism and some of the SCAs, are more slowly progressive and some, like NPH and neuroleptic Parkinsonism, are readily treatable.

We believe that unlike "atypical Parkinsonism," "Parkinson-like" carries no implication regarding treatability or prognosis. That should not be the job of the disease's name. Rather, we feel that the medical professional must educate the patient and family on the specific condition in a manner appropriate to their emotional and cognitive states.

### 3.5. Summary of proposed nosological changes

Our proposal is illustrated in the Fig. 1. We propose a modest change in the terminology for Parkinsonian disorders, retiring the term "typical" as a modifier for "PD" and replacing "atypical Parkinsonian" with "Parkinson-like." We also propose confining the term "Parkinsonism" to a phenotypic syndrome and to avoid applying it in the plural form to disease entities.

As before, the "secondary Parkinsonian disorders" would include

**Table 1**
Glossary of proposed terminology.

| Existing | Proposed | Definition/comments |
|---|---|---|
| Parkinson's disease | Parkinson's disease | No change proposed |
| parkinsonism | Parkinsonism | A group of phenotypic features, not specific disease(s). The first letter should be upper case. |
| Parkinsonisms | Parkinsonian disorders | Multiple members of a group of specific disorders featuring Parkinsonism |
| Primary parkinsonism | Primary Parkinsonian disorder | Any neurodegenerative disorder featuring Parkinsonism as a major component, at least in a majority of cases |
| Secondary parkinsonism | Secondary Parkinsonian disorder | Any non-degenerative disorder featuring Parkinsonism in at least some cases or at some point in the illness |
| Atypical Parkinsonism or atypical Parkinsonian disorder | Parkinson-like disorder | Reserve "Parkinsonism" for a group of phenotypic features (see above). Reserve "Parkinson-like" for specific disorders. |

conditions that include both non-degenerative conditions like NPH, vascular Parkinsonism, and drug-induced Parkinsonism, as well as some instances of neurodegenerative conditions like AD that can include mild Parkinsonism. Again continuing current practice, "primary Parkinsonian disorders" would apply to neurodegenerative conditions that feature Parkinsonism as a major component and have unclear causation, except for a few entities with known toxic or Mendelian genetic etiology. Refer to Table 1 for a summary of the current use of terms related to Parkinsonism and some recommendations.

### 3.6. Capitalization and apostrophes

As versions of a person's name, "Parkinson's" and "Parkinsonism" should both be capitalized, like Freudianism, Darwinism, and Marxism, not to mention the Jacksonian march, the Cushingoid facies, and the Oslerian tradition. In recent years, avoiding the possessive form of single-name eponyms has become a bit more popular outside the UK, but we see no rationale for it and feel that the possessive form better honors the individual's achievement.

## 4. Conclusions

We propose these modest changes to help clarify communication among physicians and caregivers, to reduce the stigma and stress of being considered "atypical," and to improve efficiency of research referrals. We predict that any inconvenience of the transition would be justified by these improvements in neurological practice.

## CRediT authorship contribution statement

**Junaid H. Siddiqui:** Writing – review & editing, Writing – original draft, Conceptualization. **Lawrence I. Golbe:** Writing – review & editing, Conceptualization.

## Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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