

Cambridge University Press & Assessment
978-0-521-11197-3 — Handbook of Atypical Parkinsonism
Edited by Carlo Colosimo, David E. Riley, Gregor K. Wenning
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Handbook of Atypical Parkinsonism

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Preface

This defining handbook of atypical forms of parkinsonism continues a journey towards the understanding of pathogenesis, and ultimately the cure, of these rare, largely sporadic and universal diseases of late life.

It is a journey begun 50 years ago in Toronto by neurologist J Clifford Richardson, who recognized an unusual constellation of neurological symptoms in a friend. As Richardson puzzled about its features of progressive supranuclear palsy of gaze and bulbar muscles, axial dystonia, gait impairment and dementia, he identified 3 other patients with similar symptoms and realized their illness was an unrecognized neurodegenerative syndrome. He resisted opinions by neuropathologist colleagues that it was a variant of postencephalitic parkinsonism, and in 1962 he asked Jerzy Olszewski, the new professor of neuropathology at the Banting Institute, and me as his resident to examine the seven cases that were by then identified. We found the histopathology of neurofibrillary degeneration and gliosis in brain-stem and subcortical nuclei was quite as distinctive as the clinical syndrome, but we could not be certain if it was a primary neurodegenerative disease like Alzheimer's, or an infection akin to scrapie which was just then beginning to be described.

Interest in progressive supranuclear palsy (PSP) has expanded since our description in 1964, and during the past 45 years its features have been vigorously investigated by an increasing number of neuroscientists using new biological techniques and studies that are facilitated by internet communication and international meetings. In 2010 we have learned PSP is a 4R tauopathy. Richardson's syndrome, as he originally described in 1963, is its principal manifestation but PSP disease also includes diverse phenotypes of isolated parkinsonism, corticobasal degeneration, pure akinesia, frontotemporal dementia and progressive nonfluent aphasia. Furthermore we have learned Richardson's syndrome is not unique to PSP disease, and it occurs also in (ALS)/Parkinsonism-dementia of Guam and in Guadeloupean parkinsonism.

As multiple system atrophy and corticobasal degeneration have been defined and studied in similar fashion to PSP, and as they are compared with the classical neurodegenerations of Parkinson's disease (PD), Alzheimer's disease (AD) and ALS, remarkable similarities are recognized between them.

All are featured by an abnormal and spreading protein that is specific to the neurodegeneration and accumulates in nerve cells and glia. Although some neurodegenerations are due to identified gene mutations, the majority are sporadic without obvious inheritance or family predisposition. Except for the 3 and 4R tauopathy of postencephalitic parkinsonism that followed encephalitis lethargica, and the spongiform encephalopathy of kuru that came after single feasts during mortuary cannibalism, there is no obvious preceding cause of these sporadic proteinopathies. Their onset is silent and asymptomatic, and it is not known if environmental exposure is single and isolated, or repeated and cumulative. It is not certain to what extent the pathogenesis could relate to genetic predisposition.

On Guam, a tropical island in the Western Pacific, the ALS/Parkinsonism-dementia complex (PDC) has held my interest for 27 years. It is a geographic isolate of a familial and long-latency polyproteinopathy that includes all the immunohistochemical proteins of all the major universal neurodegenerations. Its phenotypes are as diverse as its abnormal proteins, and include classical amyotrophic lateral sclerosis (ALS), PD, atypical

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parkinsonism with PSP and CBD, and Alzheimer-type dementia. During the past 50 years, this single disease has slowly declined by its principal phenotypes of ALS and PDC, and its age in onset has steadily increased. In 2010, classical ALS which was 100 times more common than elsewhere in 1953, no longer occurs on the island. Parkinsonism with dementia is uncommon and only 29 cases older than 70 were recently identified in a 2003 community survey of elderly Guamanians. The remarkable decline and disappearance of ALS/PDC in this distant place gives us hope that related and universal proteinopathies could end in the same way. But it is first necessary to identify its environmental cause and that remains our intention.

The challenge to Neurology in years ahead is to understand why abnormal proteins form, how they are acquired, and how each adversely affects the nervous system. We need to learn about their spread, and why the same protein can give rise to different phenotypes, and the same phenotype can be caused by different proteins. As we ask these questions, we will learn about protein metabolism and understand how we can influence and modify their abnormalities to prevent disease.

In 1964, we were not certain if PSP was a classical neurodegeneration akin to AD, or an infection due to a slow latent and temperate virus akin to scrapie. And we were aware of its similarities to postencephalitic parkinsonism. Forty-five years later we are still not certain. But we have learned that PSP and other neurodegenerations are due to the accumulation of abnormal proteins in the nervous system, and we are optimistic that future advances in understanding protein metabolism will give knowledge of pathogenesis and methods of cure.

I congratulate the editors for their fine presentation.

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