DIVISION OF PSP INTO PSP-P AND RICHARDSON SYNDROME IS NOT CLINICALLY USEFUL AND HELPFUL

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PSP was first described as a nosological entity in 1963 by Steele and colleagues, who gave their name to the disease. It is a multisystem neurodegenerative disorder characterized by early and progressive development of postural instability, leading to falls, typically backwards, a vertical and later horizontal supranuclear gaze palsy, pseudobulbar palsy, parkinsonism and later in the disease, a subcortical frontal – dementia.
PSP is the second most common form of Parkinsonism after Parkinson disease.
Unusual clinical findings include arm levitation, unilateral limb dystonia and compulsive repetition of movements. Apraxia has been reported and alien-limb phenomenon can be seen in some cases.
Diagnostic criteria have been formulated for possible, probable and definite PSP, with the last requiring pathological confirmation. Pathologically proven PSP has been described in patients presenting as CBD, primary progressive aphasia, FTDP-17, and Pick's disease. Vascular PSP has been described in multiinfarct state, primary phospholipids antibody syndrome, CADASIL and cerebral amyloid angiopathy. PSP-mimics include Whipple's disease, DLBD, striato-dentate-pallidal calcification, CJD, brain tumor, Gaucher's disease, neuroleptic-induced PSP, and obstructive hydrocephalus.

Diagnostic Errors: PSP Vs CBD

Subdivisions of Tauopathies are common. Clinico-pathological correlation studies have shown the clinical diagnostic accuracy is in the range of 50-60% (Gearing and colleagues, Birdi and associates). Autopsy proven corticobasal ganglionic degeneration (CBD) often presents as PSP while the corticobasal syndrome is commonly due to PSP. The diagnostic accuracy for CBD can be as low as 30% (Litvan and colleagues). PSP and CBD share important clinical, pathological, and genetic features.

PSP Vs CBD

	PSP	**CBD**
Clinical	Axial symptoms Gaze palsy Disequilibrium Early Falls	Asymmetrical movement disorder. Higher cortical dysfunction.
Pathology -Tau positive lesion	Basal ganglia and brainstem	Prefrontal and pre motor cortex, caudate nucleus, little brainstem involvement
- Typical Tau	Tufted astrocyte	Astrocytic plaque

CONFOUNDING FEATURES

1. Unilateral dystonia or apraxia can be prominent in PSP
2. S NO occurs in CBD.
3. Frontal – like syndrome occurs in both.
4. Clinical overlaps are unavoidable when either of these presents as dementia, isolated Parkinsonism or focal cortical degeneration.
5. In both tau is composed primarily of 4 repeat tau isoforms.

Subdivision of PSP

PSP has been divided into upto 8 subtypes, but the two main subtypes are PSP- RS and PSP- P (William DR, et al 2008)

	PSP – RS	**PSP – P**
Clinical	Early postural instability, falls, supranuclear gaze palsy, and cognitive dysfunction	Asymmetrical onset, tremor, moderate initial therapeutic response to levodopa.

Pathology Tau deposits	More severe and wide spread	
Disease course	short	

CONFOUNDING FEATURES

1. All cases of PSP have a subcortical concentration of pathology with deposition of 4-R tau protein in neurons and glia.
2. Severe midbrain atrophy is consistent among PSP subtypes.
3. Schiofield et al (2010) found that cortical atrophy was more severe in PSP – RS than PSP – P and affected more frontal lobe regions (Frontal pole, inferior frontal gyrus). The supra marginal gyrus was atrophic in both subtypes. However no correlations were found between the degree of atrophy and severity of tau pathology in any region assessed and the presence or absence of cardinal PSP symptoms.

So, is it CLINICALLY useful and helpful?

With clinical diagnostic accuracy being far from satisfactory even in differentiating the different tauopathies, the clinical utility of subdividing PSP is bound to be very low, given the overlapping clinical and pathological features of the subtypes and slim chances of radiological support for the subtypes. The classification is at best a pathological exercise at autopsy and is unlikely to benefit the clinician.