

Case Report

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## Progressive Supranuclear Palsy Associated with Idiopathic Normal Pressure Hydrocephalus: A Case Report and Proposal of a New Syndrome of PSP-iNPH

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### Abstract

Progressive Supranuclear Palsy (PSP) was first reported as a new syndrome and this original phenotype of PSP is called PSP-Richardson Syndrome (PSP-RS). Since then, nine different clinical phenotypes have been documented in addition to PSP-RS according to the Movement Disorder Society (MDS) criteria for PSP. However, the phenotype of PSP associated with idiopathic Normal Pressure Hydrocephalus (iNPH) has not yet been accepted, although a considerable number of patients with their combination have been reported based on pathological or clinical evidence. Herein, I report a patient presenting with PSP associated with iNPH (PSP-iNPH). Definite iNPH of this patient was diagnosed based on the Japanese diagnostic criteria for iNPH. Several disorders involving Central Nervous System (CNS) are known to coexist with iNPH and iNPH in PSP is probably induced by abnormal CerebroSpinal Fluid (CSF) dynamics due to CNS tauopathy of PSP. As the coexistence of PSP and iNPH seems to be more common than the combination of iNPH and other neurodegenerative diseases, PSP-iNPH syndrome might be accepted as a new clinical form of PSP.

**Keywords:** Progressive supranuclear palsy; Idiopathic normal pressure hydrocephalus; Tap test; Ventriculoperitoneal shunt

### Introduction

PSP was first reported in 1964 by Steel, et al. [1] as a new syndrome presenting with vertical gaze and pseudobulbar palsy, nuchal dystonia and dementia (PSP-Richardson Syndrome PSP-RS). Since then, nine different clinical phenotypes have been added to the original PSP syndrome according to the MDS criteria for PSP [2]: 1) Initial Predominance Of Ocular Motor Dysfunction (PSP-OM); 2) Postural Instability (PSP-PI); 3) Parkinsonism Resembling Idiopathic Parkinson's Disease (PSP-P); 4) Frontal Lobe Cognitive Or Behavioral Presentations (PSP-F), including behavioral variant Frontotemporal Dementia (bvFTD); 5) Progressive Gait Freezing (PSP-PGF); 6) Corticobasal Syndrome (PSP-CBS); 7) Primary Lateral Sclerosis (PSP-PLS); 8) Cerebellar Ataxia (PSP-C); and 9) Speech/Language Disorders (PSP-SL), including nonfluent/agrammatic Primary Progressive Aphasia (nfa PPA) and progressive Apraxia Of Speech (AOS) [2]. The phenotype of PSP with iNPH is not yet well accepted. A considerable number of patients have been reported who showed their coexistence based on pathological or clinical background, suggesting the presence of common pathoetiology between the two disorders [3-5]. The present case report describes a patient with clinically probable PSP-RS later developed iNPH, which initially improved after Ventriculo Peritoneal (VP) shunt placement, although the effect was not long-lasting.

### Case Report

A male patient without a remarkable medical history developed gait disturbance (small step gait), tendency to fall and mental irritability at 62 years of age. In that year, he suffered from fractures of the L3 and L4 vertebrae because of falls. When he visited our hospital at 63 years of age, he was intellectually normal, revealing a Revised Hasegawa Dementia Scale (HDS-R) of 28, indicating normal intelligence based on a full score of 30. The HDS-R is a verbal cognitive test that is as useful as the Mini-Mental State Examination (MMSE) [6]. Neurological examination revealed that his extraocular movement was slow in all directions, with restriction of 3/4 both upward and downward, which was overcome by activation with the vestibulo-ocular reflex. His range of horizontal eye movement was full. He showed no dysarthria and the deep tendon reflexes of the arms and legs were normal without positive toe extensor signs (Babinski sign). He walked with small steps in a mildly wide-based and staggering fashion. A retropulsion test was

positive. Magnetic Resonance Imaging (MRI) of the brain revealed almost normal findings for his age, with the exception of a mildly atrophic midbrain and pons (Figure 1). Therefore, the diagnosis was probable PSP with Richardson's syndrome (PSP-RS) according to the Movement Disorder Society (MDS) criteria for PSP [2].

### Results

Because his wide-based and staggering gait gradually worsened with an increase in the frequency of daily falls, follow-up brain MRI was performed at 65 years of age. This revealed increased atrophy of the midbrain and pons, especially atrophy of the rostral part of the midbrain, consistent with the hummingbird sign [7] (Figure 1). The lateral and third ventricles were enlarged. This was accompanied by narrowing of the high convexity midline subarachnoid spaces, suggesting possible iNPH, according to the Japanese diagnostic criteria for iNPH [8]. To confirm the diagnosis of iNPH, a CerebroSpinal Fluid (CSF) tap test was conducted. A 3-m Timed Up And Go Test (TUG-T) time (s), 10-m walk time (s), cadence (steps/min), step width (cm), and HDS-R were evaluated before and after lumbar puncture. The opening pressure of the CSF was 55 mmH<sub>2</sub>O and 30 mL was removed. The CSF contents were all within the normal ranges according to routine laboratory examinations. The results of the tap test are shown in Table 1, and indicate obvious improvement after the removal of CSF. Therefore, the diagnosis of "probable iNPH" was made according to the Japanese diagnostic criteria for iNPH [8]. He underwent surgical VP shunt placement, which resulted in further improvement in TUG-T. The 10-m walk and step width data did not improve in comparison to the results obtained immediately after tapping; however, they were better than the results before tapping. As a result, a diagnosis of "definite

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**Received:** 25-Mar-2026, Manuscript No. JADP-26-186913; **Editor assigned:** 27-Mar-2026, PreQC No. JADP-26-186913 (PQ); **Reviewed:** 10-Apr-2026, QC No. JADP-26-186913; **Revised:** 17-Apr-2026, Manuscript No. JADP-26-186913 (R); **Published:** 24-Apr-2026, DOI: 10.4172/2161-0460.1000655

**Citation:** Morimatsu M (2026) Progressive Supranuclear Palsy Associated with Idiopathic Normal Pressure Hydrocephalus: A Case Report and Proposal of a New Syndrome of PSP-iNPH. J Alzheimers Dis Parkinsonism 16:655

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iNPH” was made according to the Japanese diagnostic criteria for iNPH [8]. His HDS-R score remained within the normal range. At 8 months after VP shunt placement, his condition deteriorated to a state in which he could not walk. He used a wheelchair daily and showed

complete loss of extraocular movement, dysarthria, and dysphagia. His HDS-R score decreased to 24/30. He was admitted to a chronic hospital and died two years after VP shunt placement. An autopsy was not performed.

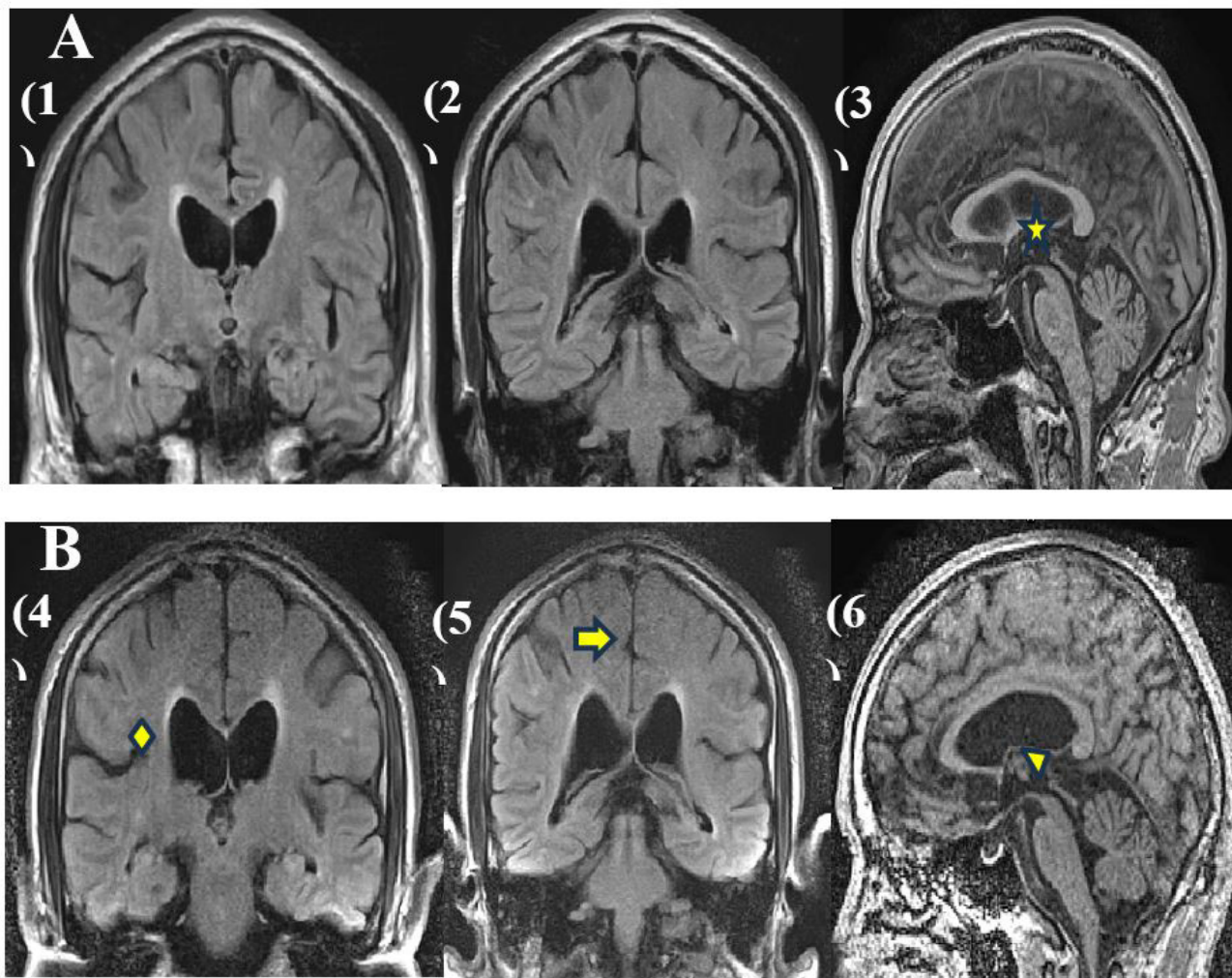


Figure 1: Brain Magnetic Resonance Imaging (MRI) of the brain.

Note: A. At 63 years of age. (1) (2) Coronal plain, (3) Sagittal plane. Mild atrophy of the midbrain and pons was observed (\*). B. At 65 years of age. (4) (5) Coronal plain, (6) Sagittal plane. The lateral ventricles were enlarged (♦), narrowing of the sulci and subarachnoid space over the high-convexity was observed (→) and obvious atrophy of the rostral part of the midbrain, showing the hummingbird sign (▼) was seen. (1) (2) (4) (5): Fluid-Attenuated Inversion Recovery (FLAIR) image, (3) (6): T1-weighted image..

	Before	After	After VP shunt
TUG-T (sec)	38.4	22.3	17.0'
10-meter walk (sec)	29.9	13.9	19.5
Cadence (steps/min)	72.3	92.0'	50.3
Step width (cm)	27.8	46.9	42.9
HDS-R	27/30	27/30	27/30

Note: TUG-T: Timed Up And Go Test (3-meter); VP: Ventriculoperitoneal; HDS-R: Revised Hasegawa Dementia Scale

Table 1: Tap test data.

## Discussion

This patient exhibited gait disturbance and repeated unprovoked falls for one year prior visiting our hospital at 63 years of age and supranuclear ophthalmoplegia was noted on the initial neurological examination. Therefore, “probable PSP-RS” was diagnosed according to the MDS clinical diagnostic criteria for PSP [2]. MRI of the brain showed mild atrophy of the midbrain, although it was not as severe as the hummingbird sign [7]. After his gait disturbance and falls worsened, MRI of the brain was performed at 65 years of age. This revealed an increase in atrophy of the midbrain, especially at the rostral part of the midbrain, consistent with the hummingbird sign. There were enlarged lateral and third ventricles accompanied by narrowing of the high convexity midline subarachnoid spaces, suggesting possible iNPH, according to the Japanese diagnostic criteria for iNPH [8]. The results of the tap test were positive and the patient underwent VP shunt placement, which resulted in further improvement in his TUG-T result.

The Japanese diagnostic criteria for iNPH [8] include “possible,” “probable,” and “definite” forms. All forms require the following conditions to be met: Possible iNPH- 1) more than one symptom of the clinical triad (i.e., gait disturbance, cognitive impairment and urinary incontinence); 2) the clinical symptoms cannot be completely explained by other neurological or non-neurological diseases; 3) no obvious preceding diseases that may be associated with ventricular dilation (including subarachnoid hemorrhage, meningitis, head injury, congenital/developmental hydrocephalus and aqueductal stenosis); probable iNPH- 1) the requirements for possible iNPH are met; 2) CSF pressure  $\leq$  200 mm H<sub>2</sub>O and normal CSF content; 3) one of a) neuroimaging features of narrowing of the sulci and subarachnoid space over the high-convexity/midline surface (Disproportionately Enlarged Subarachnoid-Space Hydrocephalus DESH) with gait disturbance (i.e., small stride, shuffle, instability during walking and increased instability on turning), b) improvement of symptoms after CSF tap test and/or a drainage test; definite iNPH- the diagnosis of definite iNPH is made when objective improvement of symptoms is observed after CSF shunt surgery. Patients in this category are considered to be “shunt responders.” iNPH must be strictly distinguished from simple ventriculomegaly associated with neurodegenerative disorders such as Alzheimer’s disease and other conditions because MRI of the iNPH brain shows narrowing of the sulci and subarachnoid space over the high-convexity/midline surface (DESH) in contrast with the expanded sulci and subarachnoid space over the high-convexity in neurodegenerative disorders. Therefore, this patient was diagnosed with “probable iNPH” after tap test and with “definite iNPH” after the CSF shunt operation.

There has been controversy regarding whether “true iNPH” can exist. Espay, et al. [9] reported that patients with so-called iNPH often (not always) coexisted with neurodegenerative disorders, such as Alzheimer’s disease, dementia with Lewy bodies and PSP. Therefore, iNPH could be a mere manifestation of neurodegenerative disorders. The Japanese diagnostic criteria for iNPH admit that many of the above-mentioned neurodegenerative disorders and cerebrovascular diseases might be associated with iNPH and that iNPH patients with coexisting neurodegenerative disorders would have a shorter clinical improvement period after shunt placement. Nevertheless, they stress that iNPH does exist and that it occurs from a unique but undetermined derangement of CSF circulation. With regard to the pathophysiology of iNPH, Wang, et al. [10] hypothesized that ventriculomegaly caused by abnormal CSF dynamics such as increased CSF pulsatility and reduced CSF drainage could initiate a vicious cycle of neurological damage, which secondarily introduces specific clinical features of iNPH. The

author speculated that these abnormal CSF dynamics would occur in some types of neurodegenerative disorders, including PSP, one of the tauopathy of the brain, causing clinical symptoms and MRI findings to be identical to those of iNPH.

Shimada, et al. [4] reported that among 85 iNPH patients, 18 (21.2%) were diagnosed as having iNPH and PSP and suggested that the coexistence of both diseases seemed to be more common than the combination of iNPH and other neurodegenerative diseases. In their series of the iNPH and PSP combination lumboperitoneal shunt surgery significantly improved the modified Rankin scale and the iNPH grading scale. Therefore, they stressed that shunt surgery could also be considered as a treatment option. Their conclusion is in good agreement with the findings observed in the author’s case.

## Conclusion

According to the MDS criteria for PSP, there are ten forms of PSP. The associated iNPH phenomenon, if present, has often been considered to be secondary ventriculomegaly due to brain atrophy caused by PSP. However, the patients with PSP associated with definite iNPH have been reported in the literature. The present report describes the case of a patient who presented with a combination of PSP and definite iNPH and based on the relevant literature dealing with the comorbidity of both disorders, PSP-iNPH syndrome might be accepted as a new clinical form of PSP.

## Acknowledgements

The author thanks the patient for his cooperation and Brian Quinn (Japan Medical Communication; <http://www.japan-mc.co.jp>) for performing the English language review.

## Ethics statement

This study was approved by the ethical committee of Tokuyama Medical Association Hospital. Informed consent was obtained from the patient for publication of this case report and accompanying images.

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