

# Progressive supranuclear palsy accompanied by spasmodic dysphonia

Nana Miyata, S.T. <sup>#1</sup>, Yuri Shingai, S.T. <sup>#1</sup>, Kumiko Kawamichi, S.T. <sup>#1</sup>, Toshio Inui, M.D. <sup>#2</sup>, Yoshiharu Arie, M.D. <sup>#2</sup>, Kazuyuki Kawamura, M.D. <sup>#2</sup>, Takao Mitsui, M.D. <sup>#2</sup>

*#1. Department of Rehabilitation, Tokushima National Hospital, National Hospital Organization, 1354 Shikiji, Kamojima, Yoshinogawa, Tokushima 776-8585 Japan.*

*#2. Department of Neurology, Tokushima National Hospital, National Hospital Organization, 1354 Shikiji, Kamojima, Yoshinogawa, Tokushima 776-8585 Japan*

Received 28 February 2016; received in received from 14 March 2016; accepted 20 March 2016

## Abstract

We report on two patients with progressive supranuclear palsy complicated by profound dysphonia. The two patients were male, 66 years old and 72 years old. We conducted sound analysis on their utterances. In the free talk, their vox hardly came out and were difficult to catch. They were able to take out the word to the lucidity in karaoke. There was no organic problem with the vocal cords in laryngendoscope testing. Interestingly, the phonation was improved by having the fiberscope inserted.

**Keywords:** PSP, stammering symptom, freezing of gait, MRI, SPECT, PSP-PAGF

## Introduction

Progressive supranuclear palsy (PSP) is a neurodegenerative extrapyramidal syndrome, characterized by motor symptoms, such as postural instability, rigidity, and akinesia. The most common problem of PSP is postural instability and frequent falls, followed by dysarthria as the second most common symptom, and bradykinesia as the third. In the late stages, swallowing difficulties, severe dysphonia and dysarthria, emotional lability, inspiratory sighs, stereotyped moaning or groaning occur. Pseudobulbar palsy with slow spastic tongue movements, reduced gag reflex, brisk jaw and facial reflexes, are common findings.

## Subjects and methods

The subjects were 12 patients with PD, and 12 patients with PSP who went to hospital for the purpose of rehabilitation. In addition

there were 12 healthy control subjects (Table 1). Utterance continuance, oral diadochokinesis and reading aloud of sentences were recorded. Sound analysis was conducted using an analysis system, AccousticCore 8 (Arcadia, Inc., Osaka, Japan). In the analysis of the utterance continuance, the longest phonation time (MPT) was measured (Figure 1A). The sound pressure level was measured every second until ten seconds after (Figure 1B). In the oral diadochokinesis analysis, the number of times that /pa/, /ta/, /ka/, /pataka/ were pronounced in five seconds was measured (Figure 1C). The sound pressure level of the first 10 words and a word at 5 seconds were measured (Figure 1D). Also we measured a long silence ward interval (Figure 1E). In the reading aloud analysis of the sentence, time was measured (Figure 1F). The prefix of each sentence and the sound pressure level of the ending of the words were measured (Figure 1F). In the present study, we assessed the cognitive function using the mini mental

scale examination (MMSE) and the frontal assessment battery (FAB). The association of cognitive function and the long silence section of oral diadochokinesis, extension of the long silence section was analyzed by Pearson's correlation coefficient test.

## Results

In the results of the cognitive function test (**Figure 2**), the MMSE did not show a significant difference between PD and PSP. The FAB significantly decreased in PSP from PD.

As to the long silence section in PD and PSP, a significant difference was found in <ta>pronunciation (**Figure 3**).

No significant correlation was found in the correlation between the long silence section and mental function scored by MMSE and FAB in Parkinson's disease (PD) and progressive supranuclear palsy (PSP) (**Figure 4**).

## Discussion

The first symptoms in two-thirds of the cases of PSP are: loss of balance, lunging forward when mobilizing, fast walking, bumping into objects or people, and falls [4]. Other common early symptoms are changes in personality, general slowing of movement,

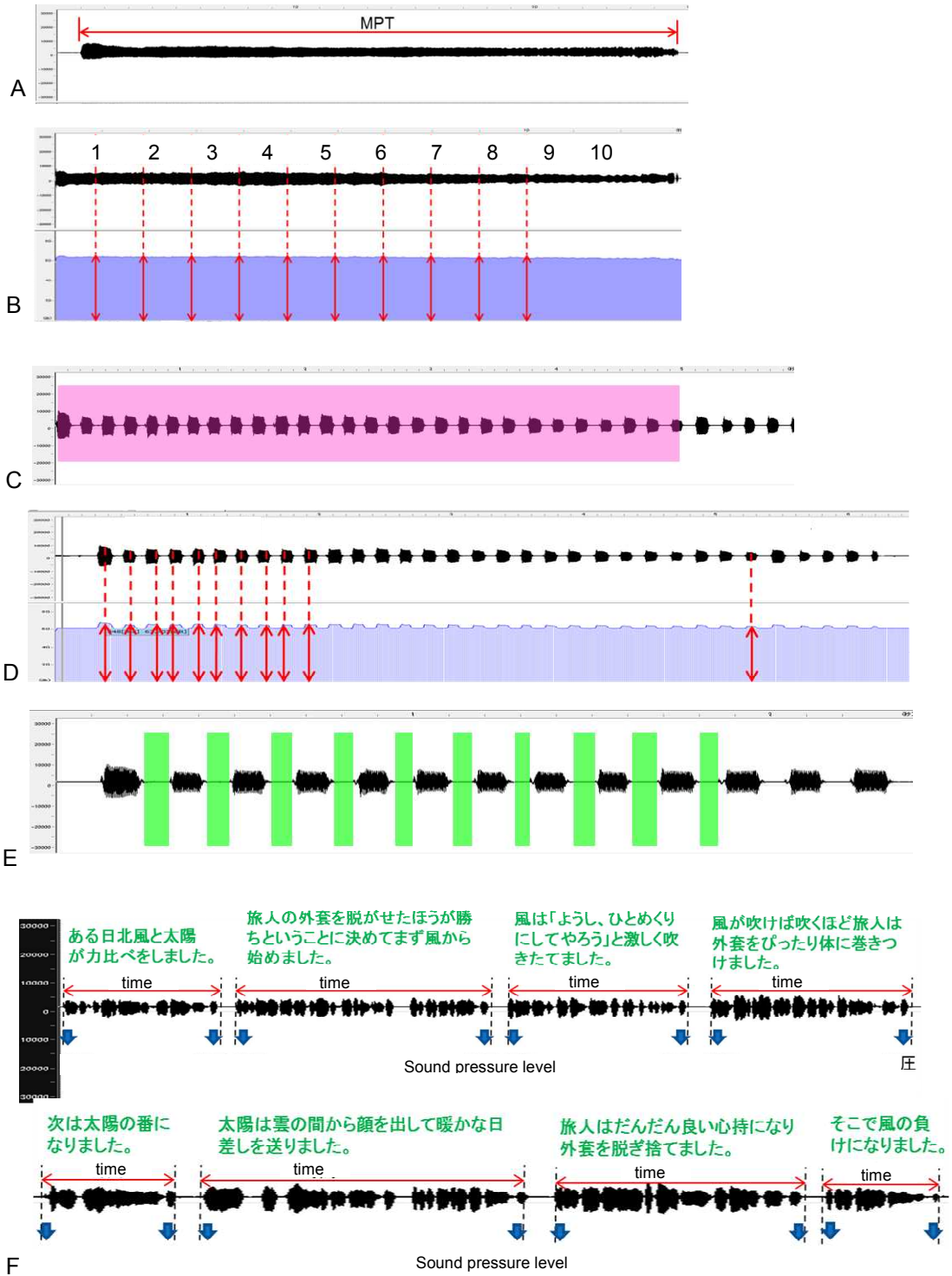
and visual symptoms. Postural instability and gait impairment are the most important disorders in the early phases of the disease. The PSP subject has a short, shuffling stepped gait, gait freezing, lurching, unsteady gait or spontaneous falls [4]. The most common problem of PSP is postural instability and frequent falls, followed by dysarthria as the second most common symptom, and bradykinesia as the third. This study will help in elucidation of the pathophysiological mechanism of PSP.

## References

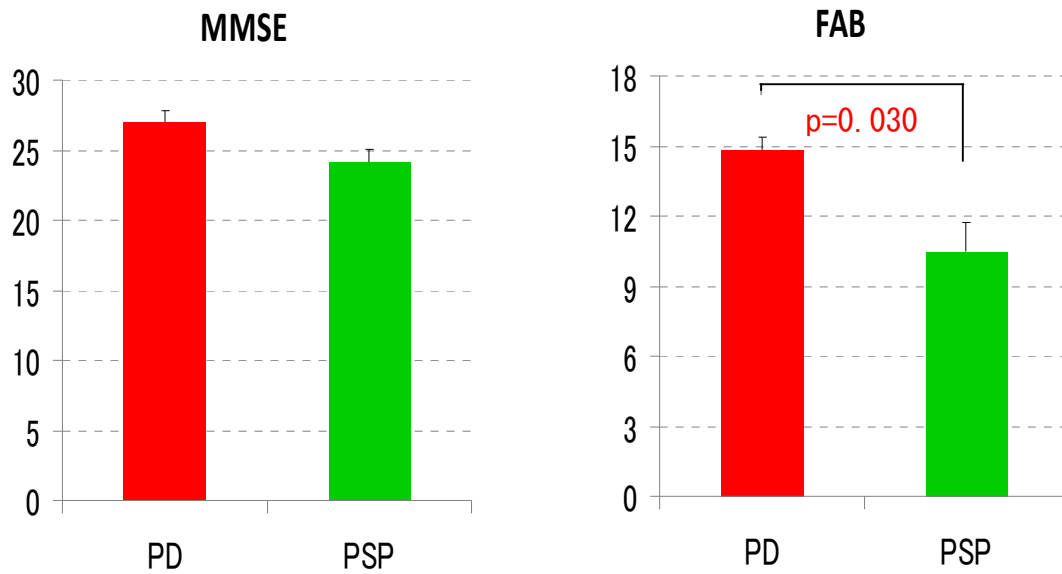
1. Williams DR, Lees AJ. Progressive supranuclear palsy: clinicopathological concepts and diagnostic challenges. *Lancet Neurol.* 2009;8(3):270-279.
2. Josephs KA, Duffy JR, Strand EA, et al. Characterizing a neurodegenerative syndrome: primary progressive apraxia of speech. *Brain.* 2012;135(Pt 5):1522-1536.
3. Josephs KA, Duffy JR, Fossett TR, et al. Fluorodeoxyglucose F18 positron emission tomography in progressive apraxia of speech and primary progressive aphasia variants. *Arch Neurol.* 2010;67(5):596-605.
4. Lubarsky M., Juncos J. L. Progressive supranuclear palsy: a current review. *Neurologist* 2008; 14, 79-88

**Table 1.** Summary of subjects

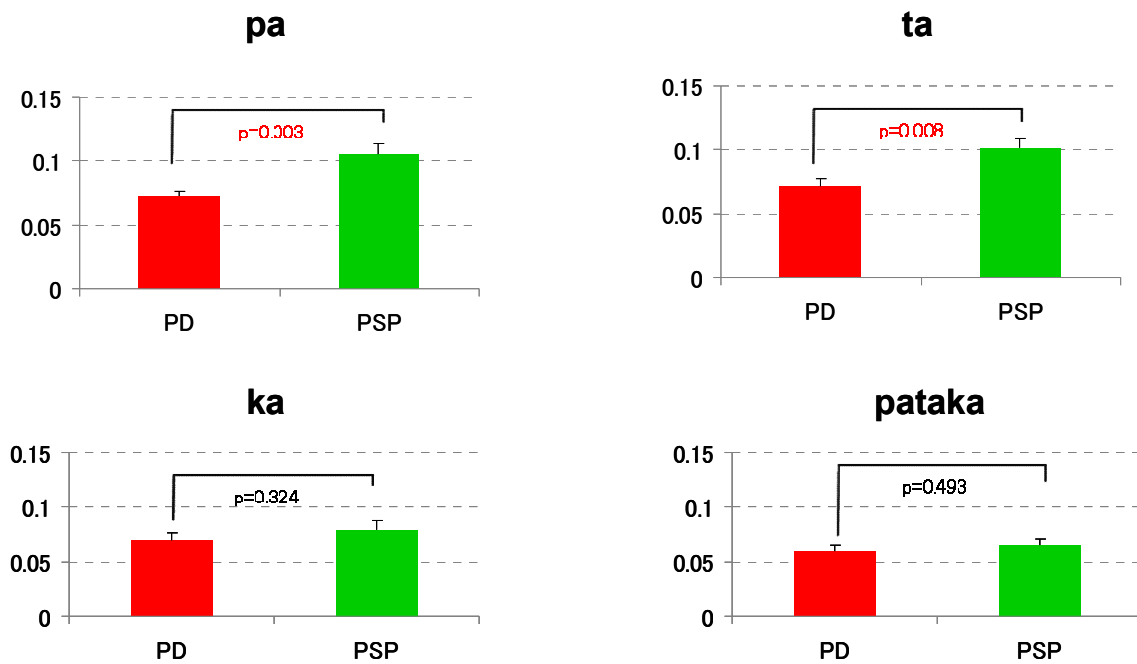
	Number	Hoehn & Yahr stage	Age	Disease duration
Parkinson's Disease	12	3.1	67.6±4.6	6.8±2.6
Progressive Supranuclear Palsy	12	-	70.6±5.3	4.5±2.9
Normal Control	12	-	-	-



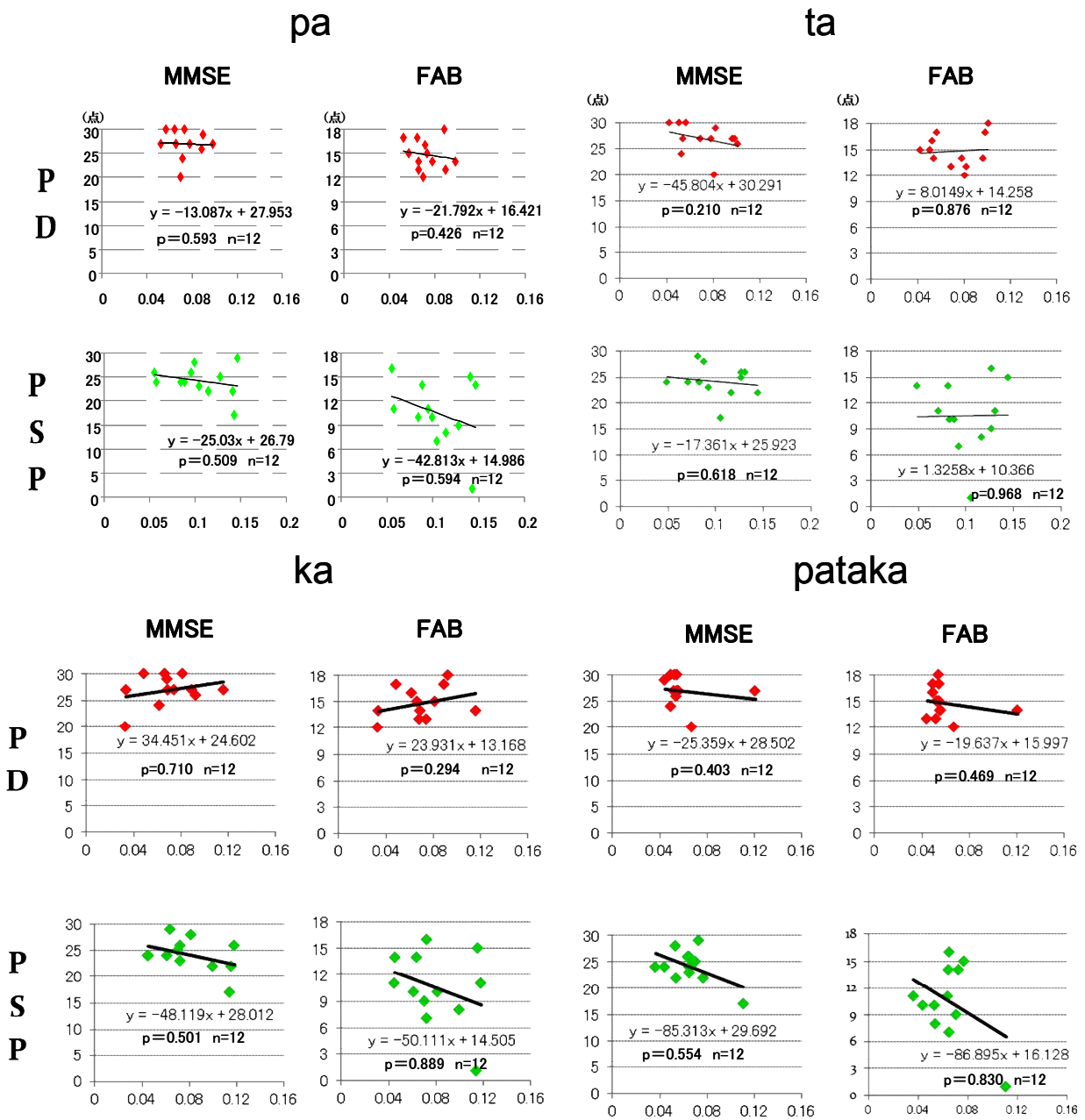
**Figure 1.** Analysis of the utterance continuance. A. Longest phonation time B. Sound pressure level every second. C. Oral diadochokinesis analysis. The number of times that /pa/, /ta/, /ka/, /pataka/ were pronounced in five seconds was measured. D. Sound pressure levels of the first 10 words and a word at 5 seconds were measured.



**Figure 2.** In the cognitive function test, MMSE did not show a significant difference between Parkinson’s disease (PD) and progressive supranuclear palsy (PSP). FAB significantly decreased in PSP from PD.



**Figure 3.** Long silence section in Parkinson’s disease (PD), progressive supranuclear palsy (PSP) and control subjects. A significant difference was found in <ta> pronunciation.



**Figure 4.** Correlation between long silence section and mental function scored by MMSE and FAB in Parkinson’s disease (PD) and progressive supranuclear palsy (PSP). No significant correlation was found.