

CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Therapy of swallowing and speech problem in patients with progressive supranuclear palsy

Mila Bunijevac

Sveti Vračevi Public Health Institution Hospital, Bijeljina, Republic of Srpska, Bosnia and Herzegovina

SUMMARY

Introduction Progressive supranuclear palsy (PSP) is a rare form of neurodegenerative extrapyramidal disease. In addition to symmetrical parkinsonism, early falls, and non-reactivity to dopaminomimetic therapy, the disease also manifests as swallowing problems with frequent choking and incomprehensible, difficult speech. In this paper, we present a case of a patient with PSP who exhibited severe swallowing and speech disorders in the clinical presentation of the disease. Appropriate therapy was applied, resulting in a positive response with partial relief of the mentioned symptoms.

Case outline A 68-year-old male patient was referred to a speech therapist by a neurologist due to difficulties with swallowing and speaking. The patient exhibited impairments in the preparatory and oral phases of swallowing, including insufficient labial occlusion and weakened tongue mobility. The patient underwent intensive speech therapy treatment for six months. The rehabilitation program led to improved swallowing function and partial improvement in speech.

Conclusion The treatment of patients with PSP should be approached seriously and interdisciplinary, given the absence of causal therapy and the reliance on symptomatic treatment for specific disabling conditions. It is essential to focus on the selection of rehabilitation programs that can improve speech and swallowing functions, as well as enhance the quality of life for patients.

Keywords: progressive supranuclear palsy; swallowing; speech; rehabilitation

INTRODUCTION

Progressive supranuclear palsy (PSP) is the second most common form of neurodegenerative parkinsonism, after idiopathic Parkinson's disease [1]. It is characterized by axial, levodopaunresponsive parkinsonism with vertical gaze palsy, early postural instability leading to backward falls, and cognitive as well as behavioral changes [2]. Nevertheless, PSP is approximately 10 times less common than typical Parkinson's disease. Some studies point out that annual prevalence is five to seven cases per 100,000 population [3], while the annual incidence rate ranges between 0.16 and 2.6 per 100,000 population [4].

According to the dominant clinical features, more than 10 subtypes of PSP have been described in the literature. The most common type subtype is Richardson's syndrome, which is characterized by early the early onset of postural instability, vertical supranuclear gaze palsy, and cognitive dysfunction. It is also associated with a faster disease progression and a shorter survival time [5]. In contrast, the occurrence of other PSP subtypes is very rare [6].

The average age of onset for disease is 60–65 years, with a slight male predominance [7]. Some suggest it is evenly distributed between males and females [8]. The average life expectancy in this form of PSP is 5–8 years [2].

The diagnosis of PSP poses a significant challenge, especially in the early stages. In

recent years, substantial progress has been made in clinical practice regarding the detection of this condition.

PSP is mainly levodopa-unresponsive and, to date, no treatment strategy has proven successful. Therefore, the main treatment approach is symptomatic therapy.

We present a patient diagnosed with PSP, who exhibited pronounced swallowing and speech symptoms and underwent speech therapy rehabilitation. The implemented rehabilitation program led to improvements in swallowing function and partially enhanced speech.

CASE REPORT

The patient is a 68-year-old retired male, with no significant health issues, aside from high blood pressure. He has no known allergies and no positive family history of serious neurological disorders. The first symptoms appeared at age 62, when he noticed difficulties with walking and speaking. Over time, these symptoms progressed and, at the time of examination, at age 68, he presented with severe postural instability characterized by frequent backward falls, as well as slurred speech and difficulty swallowing.

A neurological examination revealed vertical gaze palsy and hypomimia. The patient exhibited dysarthric speech (slow, slurred, and poorly articulated) along with dysphagia. Bilateral

Received • Примљено: May 26, 2024

Revised • Ревизија: December 26, 2024

Accepted • Прихваћено: December 29, 2024

Online first: January 9, 2025

Correspondence to:

Mila BUNIJEVAC Sveti Vračevi Public Health Institution Hospital Srpske vojske 53 76300 Bijeljina Republic of Srpska Bosnia and Herzegovina bunijevacmila@yahoo.com bradykinesia and rigidity in the arms and legs were also noted. The applause test was positive. The patient's gait was slow and unstable, requiring assistance. There were no issues with sphincter control.

Basic laboratory tests were performed (including CRP, folic acid, thyroid hormones, vitamin B12, vitamin D and immunological tests). All parameters were within normal limits, except for low vitamin D levels.

Magnetic resonance imaging of the brain showed a "hummingbird" sign and a subtly elevated signal of the tegmentum and tectum (Figure 1), as well as significant atrophy from the concave edge of the mesencephalon (Figure 2).

EEG findings were normal. Holter monitoring and an EKG showed no deviations from normal. Cardiologically, only hypertension was diagnosed, and appropriate therapy was started. There were no signs of orthostatic hypotension

The patient was diagnosed with probable PSP due to the presence of mandatory criteria and key characteristics including ocular motor dysfunction and postural instability. Additionally, the MRI findings supported the diagnosis of PSP [7]. The patient continued treatment with the adjusted antiparkinsonian drugs.

Owing to his inability to control food and liquids in the oral cavity, difficulty transferring bites or sips from the oral cavity to the pharynx, frequent choking and coughing, and difficult, often incomprehensible speech, the patient was referred for rehabilitation of speech and swallowing.

Assessment of swallowing and speech abilities

To evaluate swallowing and speech, we used the Water Swallowing Test [9], the Oral Practice Test [10], and the Global Articulation Test [11].

Water Swallowing Test is a minimally invasive screening procedure used to identify oropharyngeal dysphagia. The patient is asked to sit and drink 30 ml of room-temperature water from a glass in his hand as he normally would. During the drinking process, we record the time taken to empty the glass and the drinking profile, which may indicate the need for further investigations. The profiles are as follows:

Profile 1 – the patient can swallow all the water in one gulp without choking;

Profile 2 – the patient can drink all the water in two or more sips without choking;

Profile 3 – the patient can drink all the water in one gulp with occasional choking;

Profile 4 – the patient can drink all the water in two or more sips with occasional choking;

Profile 5 – the patient frequently chokes and finds it difficult to drink all the water.

The Oral Practice Test assesses the voluntary muscle activity of the speech apparatus including the lips, cheeks, tongue, soft and hard palate and jaw. It evaluates the ability to perform specific voluntary movements of the orofacial muscles, which are controlled by certain cranial nerves (V, VII, IX, and XII). A dysfunction in any of these cranial

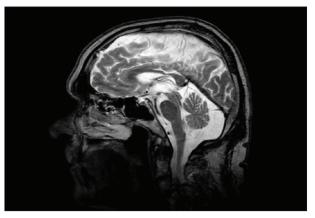


Figure 1. Magnetic resonance on the sagittal tomogram shows the so-called hummingbird sign and discretely elevated tegmentum and tectum signal; a lower ratio of the diameters of the pons and midbrain is present, as one of the differentiation factors in relation to Parkinson's disease

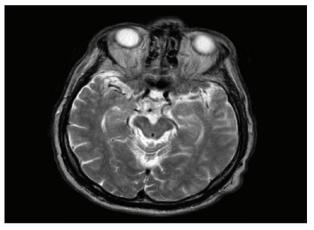


Figure 2. Magnetic resonance imaging shows significant atrophy with concave edges of the mesencephalon

nerves can lead to an impairment of the orofacial region, resulting in impaired articulation. The test consists of 21 tasks, ranging from simple actions (e.g., breathing in and out through the nose, extinguishing a match) to more complex tongue movements (e.g., creating a palatolingual groove). The subject is asked to perform each oral-motor pattern demonstrated by the examiner. For each successfully completed task, the subject receives one point, while no points are awarded for unsuccessful attempts.

The Global Articulation Test consists of 30 words and provides a detailed analysis of sounds, identifying both pathological sounds and those correctly pronounced. Correctly pronounced sounds are marked with a "+," distorted sounds with a "+," and omitted sounds with a "-."

The patient attended the speech therapy clinic twice a week for the first month, and once a week for the following five months. His daughter and wife were trained to help him carry out the exercises during daily activities. During therapy, we implemented oral practice exercises, voice articulation exercises using vowels in words and short sentences, as well as swallowing exercises to enhance the strength and mobility of the muscles essential for swallowing.

80 Bunijevac M.

Outcome of applied treatment

Table 1 presents the results of the Water Swallowing Test and the Oral Practice Test conducted before speech therapy. In the Water Swallowing Test, which lasted 15 minutes, the patient obtained Profile 5 – he frequently choked and found it difficult to finish the water.

Table 1. Results of the Water Swallowing Test and oral practice before and after speech therapy

Test	Before therapy	After therapy
Water Swallowing Test	Profile 5	Profile 2
Oral practice test (task successfully completed / number of tasks)	3/21	6/21

After six months of continuous speech therapy, we reevaluated the patient using the Water Swallowing Test, which lasted five minutes. He achieved Profile 2, meaning he could drink the water in two or more sips without choking. The patient experienced no choking episodes, as he continued to use the supraglottic swallowing technique.

On the 21-item Oral Practice Test, conducted before speech therapy, the patient scored three points for successfully completing three tasks, while he was unsuccessful in 18 tasks. Our results from an Oral Practice Test not previously administered to PSP patients, indicate notable weakness of the orofacial musculature. This reflects an inability to exert voluntary control, which is associated with impaired planning and programming of movements for speech, as well as chewing and swallowing food.

In the Oral Practice Test conducted after six months, the patient scored six points for successfully completing six tasks, while he was unsuccessful in 15 tasks. The results indicate minimal improvement, insufficient to significantly enhance the motor abilities of the oral structures, which is in line with other studies.

In Table 2, the Global Articulation Test conducted before speech therapy indicated impairment to all 30 phonemes, manifesting as distortion (9/ \pm), substitution (15/ \pm), and omission (6/-). This was especially pronounced in spontaneous speech, making it difficult to understand.

Table 2. Results of the Global Articulation Test before and after therapy

Time	Distorted and substitution voices	Omitted voices
Before therapy	24/30	6/30
After therapy	24/30	6/30

After six months, the patient distorted 10 phonemes (\pm) , substituted 14 (\pm) , and omitted 6 (-), indicating no major change in the patient's overall speech intelligibility.

The ethical commission of the Sveti Vračevi Public Health Institution Hospital in Bijeljina approved the study and consent was obtained from the patient and his family members. The research was conducted in accordance with the Declaration of Helsinki.

DISCUSSION

We present a patient diagnosed with PSP, who displayed pronounced swallowing and speech symptoms, and underwent speech therapy rehabilitation.

As the disease progresses, patients with PSP develop difficulties with swallowing and speech [12]. Dysphagia is the third most common symptom reported by PSP patients, with patients reporting a swallowing disorder three times more often than a speech disorder [13]. Some authors indicate that dysphagia in PSP occurs approximately three to four years after the onset of symptoms [14], which coincides with our patient's experience. In PSP, the oral/preparatory and oral phases of swallowing are most frequently impaired [15]. In our patient's manifestation, this likewise occurred. During the Water Swallowing Test, which lasted 15 minutes, the patient received Profile 5, reflecting frequent choking and difficulty finishing the water. The patient experienced issues with bolus preparation and transport, attributed to weakened labial occlusion, and restricted tongue mobility - essential for initiating swallowing and moving food across the base of the tongue, resulting in frequent choking.

In the treatment of patients with dysphagia, both direct and medical procedures are applied. In this case study, we utilized direct procedures (including some of the exercises for the preparation and oral phase of swallowing) along with oral motor exercises. Additionally, we implemented a supraglottic swallowing maneuver, a technique that helps reduce or control aspiration during the oral phase of swallowing, to voluntarily protect the airway.

The treatment of these patients involves exercises aimed at improving the coordination of the muscles necessary for swallowing and stimulating the swallowing reflex. Additionally, postural maneuvers (such as turning the head and tucking the chin position) are employed to redirect the flow of the bolus, helping to protect the airways and facilitate swallowing. Subsequently, the patient is advised to take small sips of water and bites of food, to cough and clear the throat after swallowing, and to modify the consistency of the food (soft/mixed/pureed) [16]. However, as the disease progresses and patients become unable to swallow, more invasive interventions are often required, such as a nasogastric tube or a percutaneous endoscopic gastrostomy [17].

Orofacial musculature plays an important role in feeding, chewing, swallowing, speaking, and facial expression. When oral musculature is weakened, patients often experience difficulties with chewing/swallowing food as well as speech/language difficulties [18], which is consistent with our case study.

A basic prerequisite for the correct pronunciation of sounds is a well-developed oral practice. This means that the muscles involved in speech can perform all the movements necessary for their proper articulation as well as for the process of chewing/swallowing food. In the Oral Practice Test conducted after six months, the subject scored six points for successfully performing six tasks, while he was unsuccessful in 15 tasks. The results indicate minimal improvement, which did not significantly

enhance the motor abilities of the oral structures – similar to results reported in other studies [19].

Good anatomy and mobility of the speech organs enable adequate speech. If one of these aspects is impaired to a lesser extent, it will have a negative impact on speech. Speech disorders in PSP manifest as weakness, incoordination, paralysis or paresis of the speech muscles, along with physiological impairments in speed, strength, sequencing, and accuracy of muscle movements, which is also characteristic of our case study [20]. Our patient exhibited dysarthria (slow, slurred, and poorly articulated speech) [21], characterized by reduced mobility, coordination, and precision of the orofacial musculature.

The Global Articulation Test revealed that numerous phonemes were distorted, substituted, or omitted, thus affecting overall intelligibility [22].

Good articulation is directly related to oral practice, meaning that the muscles involved in speech must perform well all the movements necessary for proper articulation. Speech rehabilitation in patients with PSP refers to exercises for the mobility of the speech organs, proper speech breathing, appropriate pace and rhythm of speech, and proper intonation of the voice. With these exercises, the functionality of the movement of the speech organs is achieved. According to the Global Articulation Test after conducted after six months, our patient's speech did not change significantly. Dysarthria remained present, along with impairments in a large number of phonemes due to distortion, substitution, and omission [22, 23], indicating the need for continued speech therapy rehabilitation.

Parkinsonism, vertical gaze palsy, postural instability accompanied by frequent backward falls and cognitive deficits can significantly impair or shorten the life expectancy of these patients. Therefore, it is essential to provide increased support to both the patients and their family members in the future [24, 25].

Swallowing and speech rehabilitation should be recommended as soon as symptoms become apparent, with the goal of preserving function and quality of life for as long as possible.

Conflict of interest: None declared.

REFERENCES

- Armstrong MJ. Progressive supranuclear palsy: An update. Curr Neurol Neurosci Rep. 2018;18(3):12.
 [DOI: 10.1007/s11910-018-0819-5] [PMID: 29455271]
- Roemer SF, Grinberg LT, Crary JF, Seeley WW, McKee AC, Kovacs GG, et al. Rainwater Charitable Foundation criteria for the neuropathologic diagnosis of progressive supranuclear palsy. Acta Neuropathol. 2022;144(4):603–14.
 [DOI: 10.1007/s00401-022-02479-4] [PMID: 35947184]
- Barer Y, Chodick G, Cohen R, Grabarnik-John M, Ye X, Zamudio J, et al. Epidemiology of progressive supranuclear palsy: Real world data from the second largest health plan in Israel. Brain Sci. 2022;12(9):1126. [DOI: 10.3390/brainsci12091126] [PMID: 36138862]
- Lyons S, Trépel D, Lynch T, Walsh R, O'Dowd S. The prevalence and incidence of progressive supranuclear palsy and corticobasal syndrome: A systematic review and meta-analysis. J Neurol. 2023;270(9):4451–65. [DOI: 10.1007/s00415-023-11791-2] [PMID: 37289323]
- Respondek G, Höglinger GU. The phenotypic spectrum of progressive supranuclear palsy. Parkinsonism Relat Disord. 2016;22(Suppl 1):S34–6. [DOI: 10.1016/j.parkreldis.2015.09.041] [PMID: 26421392]
- Batheja V, Fish M, Balar AB, Hogg JP, Lakhani DA, Khan M. Progressive supranuclear palsy: A case report and brief review of the literature. Radiol Case Rep. 2023;19(1):250–3.
 [DOI: 10.1016/j.radcr.2023.09.012] [PMID: 38028282]
- Viscidi E, Litvan I, Dam T, Juneja M, Li L, Krzywy H, et al. Clinical features of patients with progressive supranuclear palsy in a US insurance claims database. Front Neurol. 2021;12:571800. [DOI: 10.3389/fneur.2021.571800] [PMID: 34220661]
- Rowe JB, Holland N, Rittman T. Progressive supranuclear palsy: Diagnosis and management. Pract Neurol. 2021;21(5):376–83.
 [DOI: 10.1136/practneurol-2020-002794] [PMID: 34215700]
- Seki M, Yoshizawa H, Kubota M, Toi S, Kitagawa K. Swallowing impairment in patients with cerebral small vessel disease. TWMUJ. 2020;4:24–32. [DOI: 10.24488/twmuj.2019102]
- Stošljević L, Stošljević M, Odović M. Procena sposobnosti osoba sa motoričkim poremećajima, praktikum. Beograd: Big štampa; 2006.
- Kostić Đ, Vladisavljević S., Globalni artikulacioni test". U: Kostić Đ, Vladisavljević S, Popović M, urednici. Testovi za ispitivanje govora i jezika. Beograd: Zavod za udžbenike i nastavna sredstva; 1983. str. 15–7.
- 12. Iwashita Y, Umemoto G, Fujioka S, Arahata H, Dotsu Y, Oike A, et al. Factors that impact dysphagia and discontinuance of oral intake

- in patients with progressive supranuclear palsy. Front Neurol. 2023;14:1259327. [DOI: 10.3389/fneur.2023.1259327] [PMID: 37780702]
- Enver N, Borders JC, Curtis JA, Sevitz JS, Vanegas-Arroyave N, Troche MS. The role of vocal fold bowing on cough and swallowing dysfunction in progressive supranuclear palsy. Laryngoscope. 2021;131(6):1217–22. [DOI: 10.1002/lary.28885] [PMID: 32687232]
- Glinzer J, Flynn É, Tampoukari E, Harpur I, Walshe M. Dysphagia prevalence in progressive supranuclear palsy: A systematic review and meta-analysis. Dysphagia. 2024;39(6):1052–64.
 [DOI: 10.1007/s00455-023-10616-y] [PMID: 37336845]
- Del Prete E, Tommasini L, Mazzucchi S, Frosini D, Palermo G, Morganti R, et al. Connected speech in progressive supranuclear palsy: A possible role in differential diagnosis. Neurol Sci. 2021;42(4):1483–90. [DOI: 10.1007/s10072-020-04635-8] [PMID: 32851538]
- Lee JH, Kwon HY, Kwon KS, Park SH, Suh YJ, Kim JS, et al. Percutaneous endoscopic gastrostomy feeding effects in patients with neurogenic dysphagia and recurrent pneumonia. Ther Adv Respir Dis. 2021;15:1753466621992735.
 [DOI: 10.1177/1753466621992735] [PMID: 33764224]
- 17. Catricalà E, Boschi V, Cuoco S, Galiano F, Picillo M, Gobbi E, et al. The language profile of progressive supranuclear palsy. Cortex. 2019;115:294–308. [DOI: 10.1016/j.cortex.2019.02.013] [PMID: 30884283]
- Myung JH, Pyun SB. Effect of oral apraxia on dysphagia in patients with subacute stroke. Dysphagia. 2023;38(1):227–35.
 [DOI: 10.1007/s00455-022-10458-w] [PMID: 35508738]
- Lee JH, Kim GS, Yun DH, Chon J, Han YJ, Yoo SD, et al. The relationship between tongue pressure and oral dysphagia in stroke patients. Ann Rehabil Med. 2016;40(4):620–8.
 [DOI: 10.5535/arm.2016.40.4.620] [PMID: 27606268]
- Lupascu N, Lupescu IC, Caloianu I, Naftanaila F, Glogojeanu RR, Sirbu CA, et al. Imaging criteria for the diagnosis of progressive supranuclear palsy: Supportive or mandatory? Diagnostics (Basel). 2023;13(11):1967. [DOI: 10.3390/diagnostics13111967] [PMID: 37296819]
- Bluett B, Pantelyat AY, Litvan I, Ali F, Apetauerova D, Bego D, et al. Best practices in the clinical management of progressive supranuclear palsy and corticobasal syndrome: A consensus statement of the CurePSP centers of care. Front Neurol. 2021;12:694872. [DOI: 10.3389/fneur.2021.694872] [PMID: 34276544]

82 Bunijevac M.

- Claus I, Suttrup J, Muhle P, Suntrupe-Krueger S, Siemer ML, Lenze F, et al. Subtle esophageal motility alterations in parkinsonian syndromes: Synucleinopathies vs. tauopathies. Mov Disord Clin Pract. 2018;5(4):406–12. [DOI: 10.1002/mdc3.12616] [PMID: 30363410]
- 23. Parjane N, Cho S, Ash S, Cousin KAQ, Shellikeri S, Liberman M, et al. Digital speech analysis in progressive supranuclear palsy and corticobasal syndromes. J Alzheimers Dis. 2021;82(1):33–45. [DOI: 10.3233/JAD-201132] [PMID: 34219738]
- Respondek G, Breslow D, Amirghiasvand C, Ghosh B, Bergmans B, Van Wyk L, et al. The lived experience of people with progressive supranuclear palsy and their caregivers. Neurol Ther. 2023;12(1):229–47. [DOI: 10.1007/s40120-022-00420-1] [PMID: 36447110]
- Popović Z, Đuričković M, Ljaljević A, Matijević S, Obradović-Đuričić K. Assessment of reliability and validity of Montenegrin version of the oral health impact profile for use among the elderly in Montenegro. Srp Arh Celok Lek. 2019;147(9–10):534–40. [DOI: 10.2298/SARH180528049P]

Терапија проблема гутања и говора код болесника са прогресивном супрануклеарном парализом

Мила Бунијевац

Јавна здравствена установа Болница "Свети Врачеви", Бијељина, Република Српска, Босна и Херцеговина

САЖЕТАК

Увод Прогресивна супрануклеарна парализа је редак облик неуродегенеративне екстрапирамидне болести. Поред симетричног паркинсонизма, раних падова, нереактивности на допаминомиметичку терапију, болест се манифестује и проблемима гутања са честим загрцњивањем и неразумљивим, отежаним говором. У овом раду приказујемо болесника са прогресивном супрануклеарном парализом који у клиничкој презентацији болести има тежак поремећај гутања и говора. Примењена је одговарајућа терапија, која је резултовала позитивним одговором са делимичним олакшањем наведених симптома.

Приказ болесника Болесник мушког пола, старости 68 година, упућен је логопеду од стране неуролога због тешкоћа са гутањем и говором. Код болесника је била нарушена при-

премна и орална фаза гутања, са недовољном лабијалном оклузијом и ослабљеном покретљивошћу језика. Болесник је био укључен у интензиван логопедски третман у трајању од шест месеци. Примењени рехабилитациони програм резултирао је побољшању функције гутања и делимичном побољшању говора.

Закључак Лечењу болесника са прогресивном супрануклеарном парализом треба приступити озбиљно и интердисциплинарно, с обзиром на то да не постоји каузална терапија, те да је једина опција симптоматско лечење специфичних онеспособљавајућих стања. Неопходно је обратити пажњу на одабир рехабилитацијских програма који могу побољшати функције говора и гутања и квалитет живота болесника. Кључне речи: прогресивна супрануклеарна парализа; гутање; говор; рехабилитација