

Pearls & Oy-sters: Marionette Walk in Parkinson Disease

A Rare Dyskinetic-Dystonic Gait Pattern Complication Improved by Visual Cueing

Micol Avenali, MD, PhD,* Gloria Vaghi, MD,* Roberto De Icco, MD, PhD, Valentina Grillo, BSc, Christopher Susca, BSc, Carla Giudice, BSc, Alessia Putorti, MD, Ruggero Barnabei, MD, Marta Allena, MD, and Cristina Tassorelli, MD, PhD

Correspondence

Dr. Avenali
micol.avenali@mondino.it

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Abstract

We report a case of a 60-year-old patient with a 10-year history of Parkinson disease who developed a dyskinetic-dystonic gait pattern highly affecting his personal and social life. After multiple unsuccessful attempts to improve the clinical condition by adapting the pharmacologic treatment, the patient underwent gait rehabilitation based on the use of visual cueing. This approach induced a relevant improvement in the dyskinetic-dystonic gait. Our case contributes to the phenotypic description of motor fluctuations in advanced Parkinson disease and suggests an additional therapeutic option to mitigate their impact on motor performances.

Pearls

- Marionette walk, known as *Silly walk*, is a rare extremely disabling dyskinetic-dystonic gait pattern in Parkinson disease (PD).
- Conservative rehabilitative interventions, such as visual cues, may provide a therapeutic benefit in the dyskinetic-dystonic gait.

Oy-sters

- Marionette walk can have repercussions on a patient's social life and may represent a diagnostic challenge in clinical evaluation.
- Misdiagnosis as a psychogenic movement disorder increases the stigma associated with PD.
- Low pattern variability and the absence of distractibility are suggestive of an organic origin.
- Early diagnosis is essential for a prompt adoption of corrective measures to improve motor ability and to alleviate the disease burden.

Case Report

A 60-year-old White man developed his first Parkinson disease (PD) symptoms at the age of 51 years, with subtle resting tremor in the left upper limb, further spreading to the ipsilateral lower limb. The diagnosis of PD was supported by a DaT-SPECT scan with ((123)I)ioflupane (reduced uptake in the right putamen/caudate). His family history was negative for PD, as were other relevant amnesic comorbidities (details in eTable 1, links.lww.com/WNL/C50). Genetic screening using next-generation sequencing–based analysis was uninformative.

A year after diagnosis, he started levodopa (LD) with a daily LD-equivalent dose of 200 mg obtaining optimal control of motor symptoms, maintained after 5 years by adding entacapone.

*These authors contributed equally as first authors.

From the Department of Brain and Behavioral Sciences (M. Avenali, G.V., R.D.I., A.P., R.B., C.T.), University of Pavia; and Neurorehabilitation Unit (M. Avenali, G.V., R.D.I., V.G., C.S., C.G., A.P., R.B., M. Allena, C.T.), IRCCS Mondino Foundation, Pavia, Italy.

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After 9 years from disease onset, motor fluctuations appeared, characterized by typical peak dose dyskinesias, occasionally associated with right lower limb and foot dystonia and disabling tremor-akinetic off periods.

One year later, the patient noticed occasional and random bizarre gait characterized by involuntary dystonic movements in the lower limbs, predominantly on the left side, lasting approximately 30 minutes. Later, he started to randomly alternate “partial off” phases (characterized by sporadic gait freezing, moderate bradykinesia, left upper limb stiffness, resting, and postural tremor) with periods of dyskinetic movements, predominantly in the lower limbs, leading to what the patient described as a “marching gait.”

After failure of pharmacologic optimization in the outpatient setting, the patient was hospitalized for therapeutic evaluation and intensive rehabilitation training (details in supplementary files, links.lww.com/WNL/C50) at the Neurorehabilitation Unit of the IRCCS Mondino Foundation. At admission, MRI was unremarkable. Cognitive tests detected a multiple domain (mnestic, frontal function, and attentive) mild cognitive impairment.

As the temporal profile of gait abnormalities was not clearly associated with the intake of antiparkinsonian drugs, the patient underwent an LD challenge test after an 18-hour withdrawal of dopaminergic drugs. After 60 minutes from 150 mg of oral LD intake, tremor, rigidity, and bradykinesia disappeared. However, the improved motor performance rapidly deteriorated because of the appearance of dyskinetic movements of the lower limbs that severely impaired gait. We observed a stereotyped gait characterized by a longer swing phase, excessive elevation of the knees followed by swift extension of the legs, mimicking a kick in the air, and a prolonged heel strike phase without a proper flatfoot phase, constituting a stereotyped “marionette walk” described by some authors also as “Silly walk.”¹ The kicking pattern was more marked on the right side and was associated with a dystonic posture of the upper limbs. Three hours after LD administration, the dyskinetic-dystonic pattern disappeared, and the patient presented a global improvement in motor features. A subsequent drug challenge test with placebo in a single-blind modality failed to induce any clinical change, helping ruling out a psychogenic gait disorder.

To evaluate the potential efficacy of conservative training methods² on Silly walk, we performed a treatment approach by means of distinct visual and acoustic cues.

Administration of visual cues during the dyskinetic-dystonic gait induced a marked improvement, whereas no substantial effect was observed using acoustic cues. We therefore devised a personalized rehabilitative approach based on daily sessions of gait training using visual cues. These sessions

were also integrated with relaxation and equilibrium exercises for a total daily duration of 120 minutes, 6 days per week for 6 weeks.

The offline (chronic) efficacy of rehabilitation was evaluated instrumentally through a gait analysis system (details in the eMethods, links.lww.com/WNL/C50) at baseline and at the end of the 6-week training period.

Gait analysis without cueing recorded after the rehabilitation period confirmed a clear improvement (normalization of the difference in the stance and swing phase between left and right legs and reduction of pelvic rotation; Table 1 and Figure) that persisted at home and was still successfully transferred into daily life activities at the 2-month follow-up. EMG gait analysis also showed a marked reduction of bilateral anterior tibialis and soleus muscle coactivation during the stance phase, a pathologic condition recorded at baseline.

Videos 1–4 showing the ON/OFF phase and gait at baseline and at the end of the rehabilitation protocol are available as supplementary files (links.lww.com/WNL/C50).

Discussion

LD-induced dyskinesias represent a disabling complication in patients with PD. Dyskinesias typically appear at the peak of LD plasma levels and can be occasionally associated with dystonia, thus resulting in peculiar and misleading clinical patterns. Marionette walk is a rare side effect of long-term levodopa intake observed in early-onset PD, especially in patients with an underlying genetic pathology.³ It is characterized by stereotyped gait, usually with high knee elevation and kicking, and a possible dystonic posture of the contralateral foot and ipsilateral arm.^{1,2}

Our case contributes to the phenotypic description of this unusual gait modality that may manifest within the protean motor disturbances of advanced PD. We also provide information on a treatment option that enriches the limited therapeutic armamentarium available for these complications, when pharmacologic optimization fails.

Because of the rarity of this condition and its bizarre manifestations, in the diagnostic process it is crucial to rule out a psychogenic movement disorder. The erratic temporal pattern and the incongruity in its presentation could indeed be evocative of a functional origin.⁴

However, some features clearly point toward an organic nature: (1) the stereotyped presentation, with a low pattern variability and (2) the absence of distractibility when performing a dual task or when walking backward.^{4,5} The organic nature was ultimately confirmed by the lack of suggestibility observed during the placebo challenge test.

Table 1 Effects of Visual Cueing: Acute Effect Evaluated Without and With Cues; Chronic Effect Evaluated Offline at the End of the Rehabilitation Period

	Baseline				End of treatment	
	Without visual cues		With visual cues		Right	Left
Kinematic analysis						
Cadence (step/min) (n.v. 114 ± 4.2)	106.5 ± 1.7		114.4 ± 4		106.8 ± 4.5	
Symmetry index (%) (n.v. <3.6)	3.9		1.4		1.8	
	Right	Left	Right	Left	Right	Left
Stance phase (% step) (n.v. 59 ± 2)	56.5 ± 4.2	60.5 ± 2.6	60.6 ± 3.9	62 ± 2.9	58 ± 3.7	59.7 ± 2.1
Swing phase (% step) (n.v. 40 ± 3.6)	43.5 ± 4.2	39.5 ± 2.6	39.4 ± 3.9	38 ± 2.9	42 ± 3.7	40.3 ± 2.1
Stance duration (s) (n.v. 0.65 ± 0.07)	0.63 ± 0.05	0.68 ± 0.04	0.64 ± 0.05	0.65 ± 0.03	0.66 ± 0.07	0.67 ± 0.02
Single support phase (%) (n.v. 38.9 ± 2.6)	40.2 ± 2.7	42.9 ± 4.6	38.2 ± 3	39.3 ± 4.6	40.3 ± 4.3	42.2 ± 4.6
Double support phase (%) (n.v. 10.3 ± 3.1)	8.7 ± 3.7	7.5 ± 0.9	12.1 ± 3.4	10.3 ± 3.4	9.9 ± 4.4	7.8 ± 0.8
Swing duration (s) (n.v. 0.44 ± 0.05)	0.49 ± 0.05	0.45 ± 0.03	0.41 ± 0.05	0.44 ± 0.04	0.47 ± 0.05	0.45 ± 0.04
Walking cycle (s) (n.v. 1.1 ± 0.09)	1.12 ± 0.03	1.13 ± 0.03	1.05 ± 0.05	1.05 ± 0.04	1.13 ± 0.09	1.12 ± 0.04
EMG analysis						
Muscle Coactivation Index Stance phase (%) (n.v. <30)	44.9	71.8	29.6	36.7	25.3	30.7
Muscle Coactivation Index Swing phase (%) (n.v. <55)	36.7	27.6	23.2	14.6	15.1	22.2

Abbreviation: n.v. = normal values.

Bold values represent the items that improved with acute and chronic exposure.

The importance of an early recognition of the disorder is essential to avoid burdening the patients with unnecessary stigma, to prescribe useless medications, as well as for the prompt adoption of targeted corrective strategies.

The most common approaches proposed so far have focused on the reduction of dopaminergic therapy in patients with an established LD intake time correlation¹ at possible expense of worsening OFF state and on surgical options, such as deep brain stimulation.

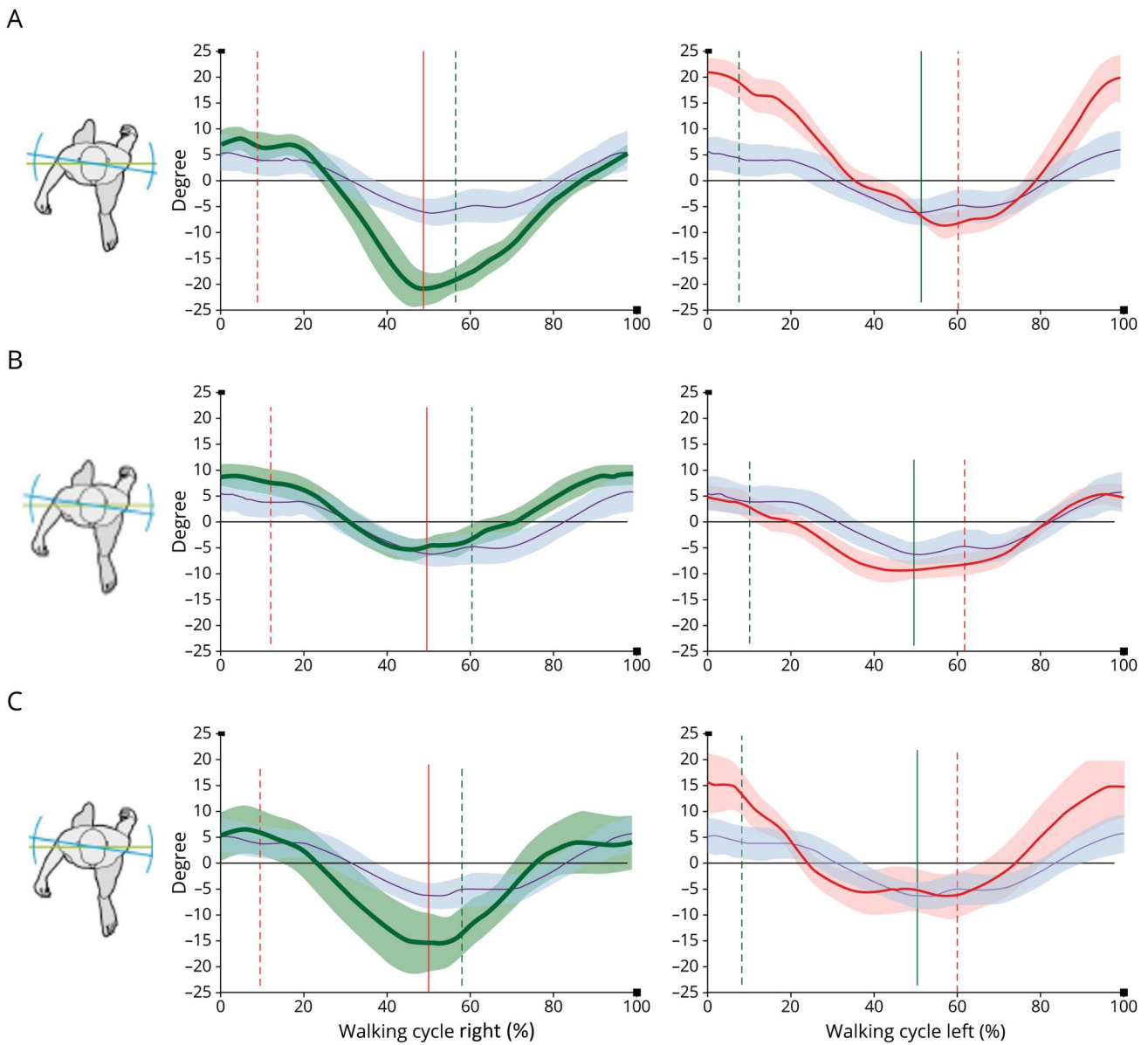
Alternative approaches have received little attention, although isolated observations suggested a possible role with a favorable risk/benefit ratio,² especially in the early phases.

In our report, we adopted an individual gait rehabilitation training based on visual cueing that induced a relevant improvement in the dyskinetic-dystonic gait. This result is not surprising when considering that neurorehabilitation positively influences neuroplasticity to modify aberrant circuits in the basal ganglia.⁶ Physical training seems to create a favorable brain environment for neuroplasticity by increasing blood flow and modulating trophic factors.⁷ Attention tasks (such as cueing strategies) act as cognitive engagement that improves neuroplasticity by triggering prefrontal circuit areas, early involved in motor learning.⁷

Thus, neurorehabilitation training may induce changes in neuronal networks that persist over time. In this frame, it is worth noting that the intensive rehabilitative approach adopted with our patient led to the retention and generalization of effect in daily life.

As previously reported, rehabilitative techniques, such as visual or auditory cues, represent valid options in the early-to-mid stage,² whereas their efficacy is attenuated in the advanced phases of disease. Visual cueing strategies enhance the so-called lateral system that includes the cerebellum and premotor and parietal cortices.⁸ This system is specifically activated during externally paced conditions, which resemble our rehabilitation cueing strategy.⁸ Auditory cues mainly modulate the impairment of internal rhythms that usually lead to festination and/or freezing of gait,⁹ acting through a facilitatory effect in spinal motor neurons.¹⁰

Our patient, in contrast to Schaeffer's description,² only responded to visual cues. The occurrence of a preferential response to one type of cueing has been previously reported,¹¹ with the identification of different cueing strategies exerting distinct effects on the PD gait pattern.¹⁰ Indeed, although acoustic cues mainly modulated cadence and speed of gait, visual cues led to an improvement in stride length, accompanied by a normalization of the stance/swing ratio.^{12,13} Thus, it is possible that in our patient visual cueing positively



(A) Pelvic rotation recorded at baseline during gait without visual cue. (B) Pelvic rotation recorded at baseline during gait under visual cue condition, showing a pattern closer to normal values (represented by the gray area). (C) Pelvic rotation recorded during gait without visual cue at the end of the rehabilitation training. In green: right foot stance phase; in red: left foot stance phase; gray areas: range of normal values. All panels show the degree of pelvic rotation (the X axis reports the percentage of the walking cycle, whereas the Y axis represents the degree of rotation).

influenced cerebellar circuit visuomotor control, with a consequent improvement on stride length and stance/swing percentage distribution.¹¹

Neurorehabilitation methods should be tailored to the single patient, using gait analysis as a valuable tool to select the most appropriate cues and to detect and document gait improvement. Patients should be encouraged to fully complete the intensive rehabilitative cycle with perseverance and, when possible, to continue training at home for a long-lasting benefit.

In conclusion, this report highlights the importance of early recognition of unusual, but highly disabling, gait patterns to

promptly adopt targeted approaches to improve motor ability and decrease the disease burden. Further studies on the use of cueing strategies specifically applied in patients with dyskinetic gait are needed to better understand the underlying mechanisms and define the criteria to identify the most effective management.

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Appendix Authors

Name	Location	Contribution
Micol Avenali, MD, PhD	Department of Brain and Behavioral Sciences, University of Pavia; Neurorehabilitation Unit, IRCCS Mondino Foundation, Pavia, Italy	Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data
Gloria Vaghi, MD	Department of Brain and Behavioral Sciences, University of Pavia; Neurorehabilitation Unit, IRCCS Mondino Foundation, Pavia, Italy	Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; analysis or interpretation of data
Roberto De Icco, MD, PhD	Department of Brain and Behavioral Sciences, University of Pavia; Neurorehabilitation Unit, IRCCS Mondino Foundation, Pavia, Italy	Drafting/revision of the manuscript for content, including medical writing for content, and analysis or interpretation of data
Valentina Grillo, BSc	Neurorehabilitation Unit, IRCCS Mondino Foundation, Pavia, Italy	Major role in the acquisition of data
Cristopher Susca, BSc	Neurorehabilitation Unit, IRCCS Mondino Foundation, Pavia, Italy	Major role in the acquisition of data
Carla Giudice, BSc	Neurorehabilitation Unit, IRCCS Mondino Foundation, Pavia, Italy	Drafting/revision of the manuscript for content, including medical writing for content
Alessia Putorti, MD	Department of Brain and Behavioral Sciences, University of Pavia; Neurorehabilitation Unit, IRCCS Mondino Foundation, Pavia, Italy	Drafting/revision of the manuscript for content, including medical writing for content, and analysis or interpretation of data

Appendix (continued)

Name	Location	Contribution
Ruggero Barnabei, MD	Department of Brain and Behavioral Sciences, University of Pavia; Neurorehabilitation Unit, IRCCS Mondino Foundation, Pavia, Italy	Major role in the acquisition of data
Marta Allena, MD	Neurorehabilitation Unit, IRCCS Mondino Foundation, Pavia, Italy	Drafting/revision of the manuscript for content, including medical writing for content
Cristina Tassorelli, MD, PhD	Department of Brain and Behavioral Sciences, University of Pavia; Neurorehabilitation Unit, IRCCS Mondino Foundation, Pavia, Italy	Drafting/revision of the manuscript for content, including medical writing for content; study concept or design; and analysis or interpretation of data

References

1. Ruzicka E, Zárubová K, Nutt JG, Bloem BR. “Silly walks” in Parkinson’s disease: unusual presentation of dopaminergic-induced dyskinesias. *Mov Disord*. 2011;26(9):1782-1784.
2. Schaeffer E, Linke G, Berg D. “Cueing” for levodopa-induced dyskinesias in Parkinson’s disease. *Front Neurol*. 2016;7:1-3.
3. Klein C, Pramstaller PP, Bernhard K, et al. Parkin deletions in a family with adult-onset, tremor-dominant parkinsonism: expanding the phenotype. *Ann Neurol*. 2000;48(1):65-71.
4. Nonnekes J, Reich SG, Bloem BR, Hallett M. Functional gait disorders: a sign-based approach. *Neurology*. 2020;94(24):1093-1099.
5. Hallett M. Functional (psychogenic) movement disorders—clinical presentations. *Park Relat Disord*. 2016;22(suppl 1):S149-S152.
6. Sudhof TC, Malenka RC. Understanding synapses: past, present, and future. *Neuron*. 2008;60(3):469-476.
7. Petzinger GM, Fisher BE, McEwen S, Beeler JA, Walsh JP, Jakowec MW. Exercise-enhanced neuroplasticity targeting motor and cognitive circuitry in Parkinson’s disease. *Lancet Neurol*. 2013;12(7):716-726.
8. Nieuwboer A, Rochester L, Müncks L, Swinnen SP. Motor learning in Parkinson’s disease: limitations and potential for rehabilitation. *Park Relat Disord*. 2009;15(suppl 3):53-58.
9. Freeman J, Cody F, Schady W. The influence of external timing cues upon the rhythm of voluntary movements in Parkinson’s disease. *J Neurol Neurosurg Psychiatry*. 1993;56(10):1078-1084.
10. Rocha PA, Porfirio GM, Ferraz HB, Trevisani VFM. Effects of external cues on gait parameters of Parkinson’s disease patients: a systematic review. *Clin Neurol Neurosurg*. 2014;124:127-134.
11. Suteerawattananon M, Morris GS, Etnyre BR, Jankovic J, Protas EJ. Effects of visual and auditory cues on gait in individuals with Parkinson’s disease. *J Neurol Sci*. 2004;219(1-2):63-69.
12. Spaulding SJ, Barber B, Colby M, Cormack B, Mick T, Jenkins ME. Cueing and gait improvement among people with Parkinson’s disease: a meta-analysis. *Arch Phys Med Rehabil*. 2013;94(3):562-570.
13. De Icco R, Tassorelli C, Berra E, Bolla M, Pacchetti C, Sandrini G. Acute and chronic effect of acoustic and visual cues on gait training in Parkinson’s disease: a randomized, controlled study. *Parkinsons Dis*. 2015;2015:978590.

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