

Ability to Cycle Despite Severe Freezing of Gait in Atypical Parkinsonism in Fahr's Syndrome



The retained ability to cycle despite freezing of gait (FOG) has been reported to be typical of patients with PD,^{1,2} and the “bicycle sign” (i.e., the loss of the ability to cycle) has been suggested as a red flag indicative of atypical parkinsonism.³ However, we present here a patient with Fahr's syndrome and severe FOG, but a remarkably preserved ability to cycle 7 years after disease onset. Fahr's syndrome encompasses a group of neurodegenerative disorders associated with calcification of the basal ganglia, cerebellum, and other brain regions.⁴ Parkinsonism with FOG and early falls are part of the clinical phenotype of this disorder.⁵

This 57-year-old patient developed difficulty walking with falls at the age of 53, followed swiftly by stuttering, erectile dysfunction, and urinary urgency. There was no relevant family history. On examination, he had hypomimia, festinant speech, echolalia, and scanning dysarthria. There was bilateral, but asymmetrical, bradykinesia and rigidity, intermittent rest tremor and bilateral postural arm tremor, dysmetria, and dysdiadochokinesia. He had marked FOG and tended to festinate backward on the pull test (see Video, Segment 1). The Mini-Mental State Examination was 25/30. A CT brain scan showed widespread calcification within the basal ganglia and dentate nuclei

consistent with Fahr's syndrome. Extensive investigation showed no other abnormalities, and a dopamine transporter (DAT) scan was normal. He was treated with levodopa (L-dopa) with a mild improvement of his symptoms. The patient reported that despite the marked FOG, he

Additional Supporting Information may be found in the online version of this article.

*Correspondence to: Maria Stamelou, Sobell Department of Motor Neuroscience and Movement Disorders, UCL Institute of Neurology, London, United Kingdom; m.stamelou@ion.ucl.ac.uk

Relevant conflicts of interest/financial disclosures: M.J.E. receives royalties from the publication of the *Oxford Specialist Handbook of Parkinson's Disease and Other Movement Disorders* (Oxford University Press, 2008) and receives research support from a National Institute for Health Research (NIHR) grant, where he is the PI. He has received support for conference travel and accommodation from Teva Pharmaceuticals and UCB Pharmaceuticals. K.B. received funding for travel from GlaxoSmithKline, Orion Corporation, Ipsen, and Merz Pharmaceuticals, LLC, serves on the editorial boards of *Movement Disorders* and *Therapeutic Advances in Neurological Disorders*, receives royalties from the publication of the *Oxford Specialist Handbook of Parkinson's Disease and Other Movement Disorders* (Oxford University Press, 2008), received speaker honoraria from GlaxoSmithKline, Ipsen, Merz Pharmaceuticals, LLC, and Sun Pharmaceutical Industries Ltd., and received research support from Ipsen and the Halley Stewart Trust through Dystonia Society UK and the Wellcome Trust MRC strategic neurodegenerative disease initiative award (Ref. no.: WT089698). M.S. and M.K. have no disclosures. Full financial disclosures and author roles may be found in the online version of this article.

Published online 28 May 2011 in Wiley Online Library (wileyonlinelibrary.com). DOI: 10.1002/mds.23794

could get around his local village by cycling with no difficulty (see Video, Segment 2).

Here, we report a patient with atypical parkinsonism and severe FOG who, nonetheless, has a negative bicycle sign, which has been suggested to distinguish patients with PD from atypical parkinsonism. This case suggests that this sign should be used with some caution in this regard. It remains unclear why, in some patients, there is a dissociation between severe difficulty with generating leg movements while walking, manifesting as FOG, and preserved ability to generate leg movements while cycling, although some possible suggestions have been made.² For example, the action of cycling might represent a type of an external pacing cue that helps to overcome freezing.⁷ Another notable aspect is that the speed of leg movements seems to improve when cycling. This is not simply an improvement related to the “on” state in PD patients, as this was also observed in our patient who had a normal DAT scan and little response to L-dopa. The improvement may be related to “paradoxical kinesia,” a brief, sudden period of mobility in response to stress or life-threatening events.⁶

The bicycle sign has been suggested as a new red flag for distinguishing PD from atypical parkinsonism.³ One issue with the previous report is that the atypical parkinsonian patients, taken together as a group, were significantly older and more impaired in terms of UPDRS, postural instability, and ataxia than PD patients. It is unclear whether patients in the earlier stages of atypical parkinsonian conditions also lose the ability to cycle. In conclusion, here, we demonstrate a patient with atypical parkinsonism and marked FOG resulting from Fahr’s syndrome with a perfectly preserved ability to cycle 7 years after disease onset. We suggest that the bicycle sign should be used with caution as a red flag to distinguish PD from atypical parkinsonism.

Legend to the Video

The first part of the video demonstrates the patient with Fahr’s syndrome and severe freezing of gait, which improves when using a visual cue stick. The second part of the video shows the patient cycling without any difficulties. ■

Acknowledgments: M.S. is supported by an EFNS scientific fellowship. This work was undertaken at UCLH/UCL, which received a proportion of funding from the Department of Health’s NIHR Biomedical Research Centers funding scheme. We thank the patient and his caregivers for their consent to publish the video.

Maria Stamelou, MD, PhD,^{1,2*} Maja Kojovic, MD,¹
Mark J. Edwards, MD, PhD,¹ and Kailash P. Bhatia, MD¹

¹*Sobell Department of Motor Neuroscience and
Movement Disorders, UCL Institute of
Neurology, London, United Kingdom; and*
²*Department of Neurology, Philipps University,
Marburg, Germany*

References

1. Snijders AH, Bloem BR. Images in clinical medicine. Cycling for freezing of gait. *N Engl J Med* 2010;362:e46.
2. Snijders AH, Toni I, Ruzicka E, Bloem BR. Bicycling breaks the ice for freezers of gait. *Mov Disord* 2011;26:367–371.
3. Aerts MB, Abdo WF, Bloem BR. The “bicycle sign” for atypical parkinsonism. *Lancet* 2011;377:125–126.
4. Oliveira JR, Spiteri E, Sobrido MJ, et al. Genetic heterogeneity in familial idiopathic basal ganglia calcification (Fahr disease). *Neurology* 2004;63:2165–2167.
5. Kostic VS, Lukic-Jecmenica M, Novakovic I, et al. Exclusion of linkage to chromosomes 14q, 2q37 and 8p21.1-q11.23 in a Serbian family with idiopathic basal ganglia calcification. *J Neurol* (in press). DOI: 10.1007/s00415-011-5985-1.
6. Bonanni L, Thomas A, Onofrij M. Paradoxical kinesia in parkinsonian patients surviving earthquake. *Mov Disord* 2010;25:1302–1304.
7. Robottom BJ, Weiner WJ, Asmus F, Huber H, Gasser T, Schols L. Kick and rush: paradoxical kinesia in Parkinson disease. *Neurology* 2009;73:328; author reply, 328–329.