Teaching Video NeuroImages: Cautious walking gait in siblings with hereditary hyperekplexia

Chao Zhang, MM,* Shi-Ge Wang, MM,* Yan Wang, MM, Xiao-Li Liu, MM, and Li Cao, MD, PhD

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Correspondence
Dr. Cao
caoli2000@yeah.net
or Dr. Liu
liuxl@rjlab.cn

A 48-year-old man and his 41-year-old sister complained of pathogenic startles since birth. They carried a homozygous deletion mutation in *GLRA1*: c.754delC (p. L252X). They felt stiff in 4 limbs with expressionless face when they were alarmed by unexpected touch and sound stimulation, startled, and fell down with stiffed limbs since walking independently. Therefore they adopted a cautious walking gait as a subconscious self-protection (videos 1 and 2).

With clonazepam treatment, pathologic cautious walking gait can be dramatically improved. Cautious walking gait is an interesting and easily observable manifestation of hereditary hyperekplexia.[1,2]

MORE ONLINE

Videos

→Teaching slides
links.lww.com/WNL/A894

Disclosure

C. Zhang, S. Wang, and Y. Wang report no disclosures relevant to the manuscript. X. Liu is in charge of Shanghai Municipal Commission of Health and Family Planning (20184Y0056). L. Cao is in charge of National Natural Science Foundation of China (81571086 and 81870889), Shanghai Municipal Education Commission-Gaofeng Clinical Medicine Grant (20161401), and Interdisciplinary Project of Shanghai Jiao Tong University (YG2016MS64). Go to Neurology.org/N for full disclosures.

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*These authors contributed equally to this work.

From the Department of Neurology (C.Z., S.-G.W., Y.W., X.-L.L., L.C.), Rui Jin Hospital & Rui Jin Hospital North, Shanghai Jiao Tong University School of Medicine; and Department of Neurology (X.-L.L.), Shanghai Fengxian District Central Hospital, Shanghai Jiao Tong University Affiliated Sixth People's Hospital South Campus, Shanghai, China.

Appendix Authors

Name	Location	Role	Contribution
Chao Zhang, MM	Department of Neurology, Rui Jin Hospital and Rui Jin Hospital North, Shanghai Jiao Tong University School of Medicine	Author	Data collection and evaluation, video shooting, drafting the manuscript
Shi-Ge Wang, MM	Department of Neurology, Rui Jin Hospital and Rui Jin Hospital North, Shanghai Jiao Tong University School of Medicine	Author	Image editing, manuscript drafting and revision
Yan Wang, MM	Department of Neurology, Rui Jin Hospital and Rui Jin Hospital North, Shanghai Jiao Tong University School of Medicine	Author	Data evaluation
Xiao-Li Liu, MM	Department of Neurology, Shanghai Fengxian District Central Hospital, Shanghai Jiao Tong University Affiliated Sixth People's Hospital South Campus	Corresponding author	Data evaluation, manuscript revision
Li Cao, MD, PhD	Department of Neurology, Rui Jin Hospital and Rui Jin Hospital North, Shanghai Jiao Tong University School of Medicine	Corresponding author	Funding, data collection and evaluation, study design and supervision, manuscript revision, final approval

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