LETTER TO THE EDITOR

Tremor in a Bassoonist: Tremor in Dystonia or Essential Tremor?

Jung E Park,1,2 Vesper Fe Marie L. Ramos,1,3 Mark Hallett1

1Human Motor Control Section, National Institute of Neurological Disorders and Stroke, National Institutes of Health, Bethesda, MD, USA
2Department of Neurology, Dongguk University Ilsan Hospital, Goyang, Korea
3FDA Center for Devices and Radiologic Health, Office of Device Evaluation, Division of Neurological and Physical Medicine, Neurodiagnostics and Neurosurgical Devices Branch, White Oak, MD, USA

Dear Editor,

Tremor is a common clinical feature in dystonia. It can be seen in both affected and unaffected body parts in patients with dystonia. Clinically, it may not be easily distinguishable from essential tremor, currently defined as a bilateral, largely symmetric postural or kinetic tremor involving the hands and forearms that is visible and persistent, with an associated or isolated head tremor in the absence of abnormal posturing.1 Efforts to differentiate these types of tremor include using neurophysiological tests such as somatosensory temporal discrimination threshold (TDT) testing, electromyography, accelerometry and reciprocal inhibition.2-3 In patients with dystonia, TDT values have been found to be prolonged, postulated to reflect abnormalities in the basal ganglia.2,4-6 On the other hand, investigations on patients diagnosed with essential tremor have revealed TDT values to be normal in those with upper limb tremor.2,7

To illustrate the use of TDT in tremor diagnosis, we report a case of tremor in a 48-year-old right-handed man. His symptoms began 12 years prior to consult, initially task-specific, beginning in the left hand while playing the bassoon. The tremor progressed to involve both hands, the head, voice and trunk, often simultaneously. Tremor and head pulling to the right would worsen with manual activities and emotional stress. He was seen in both upper extremities and trunk, as well as a vocal tremor. Right head tilt was present, most prominent on playing the bassoon. There were no sensory tricks. Bilateral arm posturing was noted with walking, greater on the right (Supplementary Video in the online-only Data Supplement).

Magnetic resonance imaging of the brain and a dopamine transporter single photon emission computed tomography scan were performed, which were both unremarkable. He did not undergo DYT gene testing. Somatosensory TDT testing was performed at our institution, with the patient appropriately focusing on the task (reporting one stimulus vs. two). The mean and range (215.83 ± 14.29 ms) of the threshold for bilateral upper extremities were markedly prolonged, compatible with the results seen in patients with dystonia (110 ± 31.3 ms).2

Our case illustrates a tremor in a 48-year-old man that was first noticed to occur in the left hand when playing the bassoon, eventually involving both hands, the head, voice and trunk. Differentiating tremor in dystonia from essential tremor based on clinical criteria alone can be difficult, as both types of trem-
or are often symmetric, postural and kinetic and the dystonia may not be obvious on initial examination. TDT testing may be a useful tool for this purpose, along with other neurophysiological testing methods. Prolonged thresholds have been found in various dystonia phenotypes, including cervical dystonia, writer’s cramp, blepharospasm, musician’s dystonia and spasmodic dysphonia.

The dramatically prolonged TDT results, the cervical dystonia noted on examination and the history of nearly simultaneous onset of tremor in the neck and upper extremities suggest that this is likely tremor in dystonia. We are ascribing his cognitive problems to depression, but this aspect will require continued attention.

Differentiating tremor in dystonia from essential tremor is important in considering treatment options and further testing, including genetic counseling. Thus, TDT may be another useful neurophysiological tool in making this distinction.

Supplementary Video Legends
Patient playing the bassoon. A right head tilt and bilateral postural and kinetic tremor in the upper extremities are seen. A voice tremor was also noted.

Supplementary Materials
The online-only Data Supplement is available with this article at http://dx.doi.org/10.14802/jmd.15054.

Conflicts of Interest
The authors have no financial conflicts of interest.

Acknowledgments
This work was supported by the National Institute of Neurological Disorders and Stroke (NINDS) Intramural Research Program. Dr. Park has received research grants from the Dystonia Medical Research Foundation and SK chemicals. Dr. Ramos has received a research grant from the Dystonia Medical Research Foundation. Dr. Hallett serves as Chair of the Medical Advisory Board for and receives honoraria and funding for travel from the Neuropeptide Institute. He may accrue revenue on US Patent #7,407,478 (Issued: August 5, 2008): Coil for Magnetic Stimulation and methods for using the same (H-coil); in relation to the latter, he has received license fee payments from the NIH (from Brainsway) for licensing of this patent. He is on the Editorial Board of 20 journals, and received royalties and/or honoraria from publishing from Cambridge University Press, Oxford University Press, John Wiley & Sons, Wolters Kluwer, Springer, and Elsevier. He has received honoraria for lecturing from Columbia University. Dr. Hallett’s research at the NIH is largely supported by the NIH Intramural Program. Supplemental research funds have been granted by the Kinetics Foundation for studies of instrumental methods to monitor Parkinson’s disease, BCN Peptides, S.A. for treatment studies of blepharospasm, Medtronics, Inc., for studies of deep brain stimulation, Parkinson Alliance for studies of eye movements in Parkinson’s disease, Merz for treatment studies of focal hand dystonia, and Allergan for studies of methods to inject botulinum toxins.

REFERENCES
1. Deuschl G, Bain P, Brin M. Consensus statement of the Movement Disorder Society on tremor. Ad Hoc Scientific Committee. Mov Disord 1998;13 Suppl 3:2-23.
2. Tinazzi M, Fasano A, Di Matteo A, Conte A, Bove F, Bovi T, et al. Temporal discrimination in patients with dystonia and tremor and patients with essential tremor. Neurology 2013;80:76-84.
3. Münchau A, Schrag A, Chuang C, MacKinnon CD, Bhatia KP, Quinn NP, et al. Arm tremor in cervical dystonia differs from essential tremor and can be classified by onset age and spread of symptoms. Brain 2001;124(Pt 9):1765-1776.
4. Bradley D, Whelan R, Kimmich O, O’Riordan S, Mulrooney N, Brady P, et al. Temporal discrimination thresholds in adult-onset primary torsion dystonia: an analysis by task type and by dystonia phenotype. J Neurol 2012;259:77-82.
5. Scontrini A, Conte A, Defazio G, Fiorio M, Fabbbrini G, Suppa A, et al. Somatosensory temporal discrimination in patients with primary focal dystonia. J Neurol Neurosurg Psychiatry 2009;80:1315-1319.
6. Fiorio M, Tinazzi M, Bertolasi L, Alogli SM. Temporal processing of visuotactile and tactile stimuli in writer’s cramp. Ann Neurol 2003;53:630-635.
7. Conte A, Ferrazzano G, Manzo N, Leodori G, Fabbbrini G, Fasano A, et al. Somatosensory temporal discrimination in essential tremor and isolated head and voice tremors. Mov Disord 2015;30:822-827.