JMD
http://dx.doi.org/10.14802/jmd.15054 / J Mov Disord 2016;9(2):124-125
pISSN 2005-940X / eISSN 2093-4939

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 Hallett[1]

[1]Human Motor Control Section, National Institute of Neurological Disorders and Stroke, National Institutes of Health, Bethesda, MD, USA
[2]Department of Neurology, Dongguk University Ilsan Hospital, Goyang, Korea
[3]FDA 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 by several neurologists and was given varying diagnoses of dystonia and essential tremor. Sodium oxybate dampened the tremor, but had to be discontinued due to side effects. Current medications included propranolol extended-release 60 mg daily, topiramate 100 mg twice daily, clonazepam 1 mg twice daily and escitalopram 20 mg daily. The patient's past medical history was positive for depression, alcoholism (sober for the past eight years) and asthma. There was no family history of tremor, and both parents have a history of strokes. He is a smoker, works as a priest, is separated and lives alone.

Exam revealed mildly depressed affect, bradyphrenia and intermittent word-finding difficulty. The Montreal Cognitive Assessment score was 23/30 with errors in the following domains; executive, attention, language, and delayed recall. A high-frequency, asymmetric postural and kinetic tremor was noted 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 his attention on the task (reporting one stimulus vs. two). The mean and range (215.83 ± 14.29 ms) of the thresholds 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-

Received: November 12, 2015 **Revised:** December 6, 2015 **Accepted:** December 7, 2015
Corresponding author: Jung E Park, MD, Human Motor Control Section, National Institute of Neurological Disorders and Stroke, National Institutes of Health, Building 10, Room 7D42, 10 Center Drive, Bethesda, MD 20892, USA / **Tel:** +1-301-496-9526 / **Fax:** +1-301-480-2286 / **E-mail:** junge.park@nih.gov

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.[2] Prolonged thresholds have been found in various dystonia phenotypes, including cervical dystonia, writer's cramp, blepharospasm, musician's dystonia and spasmodic dysphonia.[2,4]

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 Neurotoxin Institute. He may accrue revenue on US Patent #6,780,413 B2 (Issued: August 24, 2004): Immunotoxin (MAB-Ricin) for the treatment of focal movement disorders, and 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.

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